Differential Diagnosis of Clival Lesions — Literature Review of the Clinical and Radiological Features

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Abstract

Background and objectives: Solitary lesions of the clivus are uncommon and represent a diagnostic and therapeutic challenge. There is a wide range of entities, with several behaviours, that can affect the central region of skull base. Imaging studies constitute an important tool in differentiating between malignant, benign and non-tumoral lesions. The aim of this study is to provide an overview of the clinical and radiological characteristics of clival lesions.

Subjects and methods: A literature review was conducted, with special attention to clinical and imaging features of the different diseases that can affect the clivus. Clinical examples of some cases are also given.

Results: Chordoma is the most frequent lesion in this location, although they are difficult to distinguish from chondrosarcomas or metastases on CT scans and MR images. Benign neoplasms can also be life-threatening and clinically undistinguishable from malignancies. Non-tumoral lesions can mimic neoplasms and lead to unnecessary aggressive treatments.

Conclusion: Clival disorders comprise a large range of entities with different characteristics and behaviors. Imaging study allows an evaluation of the tumor extent, a reliable distinction between benign and malignant processes and helps in the suspicious diagnosis. Despite imaging is an important supporting element, a final diagnosis often cannot be found without histologic biopsy.

INTRODUCTION

The skull base forms the floor of the cranial cavity and separates the brain from facial structures. This complex region is composed by five bones and can be subdivided into three regions: the anterior, middle, and posterior cranial fossae. Tumors that arise in the skull base are usually classified based on this division.

The posterior cranial fossa is composed by the occipital bone, with contributions from the sphenoid and temporal bones. The basilar part of the occipital bone and the posterior portion of the sphenoid body form the clivus, which constitutes the most anterior region of the posterior fossa. The clivus occupies a central position in the skull base, establishing close relationships with the brainstem and other noble structures [1,2].

Lesions that arise from or within the clivus are rare and represented by a wide range of different diseases. There are different studies describing neoplastic processes - like chordoma, plasmacytoma, multiple myeloma, pituitary adenoma, chondrosarcoma, meningioma and metastatic tumors - or tumor-like conditions affecting the clival region - such as fibrous dysplasia. Most of these studies, however, are limited to single case reports or case series. Revision papers on this matter are scarce.

Although most of the lesions show a benign behaviour, clival diseases can be potentially devastating and life-threatening [3,4]. These tumours usually present as incidental imaging findings. Patients may complain of headache or cranial neuropathies, especially diplopia due to abducens nerve palsy. Then, other cranial nerves or the brainstem can be affected by compression and invasion.

Given the poor clinical presentation, imaging plays a pivotal role in achieving a correct diagnosis. High resolution computed...
tomography (CT) and magnetic resonance imaging (MRI) are helpful to differentiate between non-tumoral and tumoral conditions and in searching for malignant characteristics. They also evaluate the extension of the lesions and the invasion of adjacent structures. CT is more sensitive in the assessment of trabecular and cortical bone destruction and MRI allows visualization of bone marrow and soft tissue involvement [5].

Scintigraphy permits a functional approach in the research of other bone lesions. The Fluorodeoxyglucose - Positron Emission Tomography/Computed Tomography (FDG-PET/CT) scanning technique may be helpful in the evaluation of metabolic activity of the tumor and in the investigation of other involved structures.

A correct differential diagnosis of a clival lesion is important to define the most effective therapeutic or surgical approach. For a definitive diagnosis, biopsy or resection of the lesion followed by histopathologic analyses are usually necessary. However, this task can sometimes be difficult. That is why an accurate diagnosis needs a close cooperation between surgeons, radiologists, and pathologists in a real multidisciplinary team work [6].

This study aims to review the main differential diagnosis of the lesions that typically arise from the clivus and describe the imaging characteristics that can be helpful to perform a correct diagnosis.

- Hyperintensity on T2-weighted images and strong contrast enhancement with gadolinium suggest chordoma and chondrosarcoma.
- Chordoma reacts with cytokeratin and EMA, whereas chondrosarcoma is negative for cytokeratin and EMA.
- Chordomas commonly enhance markedly with gadolinium, although EP does not show any enhancement with contrast addition.

**DISCUSSION & CONCLUSION**

**Anatomy and Embryology**

The term clivus was firstly used by von Sommerring in 1791 and it meaning “slope or declivity”. The clivus is a sloping bony structure that forms the anterior portion of the posterior cranial fossa, in a median position. It occupies a central part in the skull base and is bounded posteriorly by the margin of the foramen magnum. The anterior margin is not well defined, since it blends with the sphenoid sinus. The lateral margins are formed by the petro-occipital fissures, separating it from the petrous portion of the temporal bones. The exocranial surface of the clivus is roughened by the attachments of the fibrous raphe of the pharynx, giving rise to the pharyngeal tubercle and faces the roof of the nasopharynx. The intracranial surface is smooth and faces the pons and medulla [1,2]. The clivus is composed by the posterior portion of the body and then the dorsum sellae of the sphenoid bone, also called basisphenoid, and the basilar portion of the occipital bone, denominated basiocciput. This bony structure can be divided into upper, middle, and lower thirds. The upper third is formed by the basisphenoid bone and it is at the level of the sphenoid sinus. The middle and lower thirds are formed by the basiocciput and are divided by a line connecting the caudal ends of the petroclival fissures. Given its central position in the skull base, the clivus has close relationships with nervous and vascular structures. Lateral to the clivus is a groove for the inferior petrosal sinus, the foramen magnum and the internal carotid arteries. Superiorly, basilar artery and its branches run between the clivus and pons. The abducent nerve arises from the brainstem between the pons and medulla and passes forward, piercing the dura covering the clivus. Continuing upward in the Dorello’s canal, it crosses the superior edge of the petrous temporal bone, enters and crosses the cavernous sinus [1, 2, 7].

The clivus develops by endochondral ossification. The notochord in the cephalic region of the fetus has begun to develop by the third week of gestation. Chondrofication centres begin in the future basioocciput and spread superiority to unite with cartilaginous centres in the region of the basiosphenoid. By the sixth week of fetal life, the basal portion of the sphenoid bone contains one or two ossification centers. At birth, the clivus consists of partially ossified components of the basiocciput and sphenoid body, separated by the spheno-occipital synchondrosis. The synchondroses between the basilar and the lateral parts of the occipital component usually close very early, at between one and four years. However, the spheno-occipital synchondrosis closes between 18 to 25 years. This synchondrosis, which is one of the last sutures in the body to fuse, has an important role in the postnatally growth of the skull base [1,2].

**Normal Imaging of the Clivus**

Imaging study is determinant in detection and diagnosis of clival lesions. High resolution CT-scan and MRI are complementary techniques necessary to clarify and characterize the lesion. The clivus is best analysed in the sagittal and axial view. In a sagittal view, divus seems a triangle with the base in the dorsum sellae and the apex pointed to the foramen magnum. A normal clivus appears on the CT-scan as a central portion of cancellous bone with marrow bone elements, bounded by peripheral compact cortical bone. On the normal MRI, the signal intensity depends on the nature of the marrow in the cancellous bone. Young patients reveal a low T1 signal due to the predominance of the hematopoietic cells. With advancing age, occurs conversion of red to yellow bone marrow. With the fatty transformation of the bone marrow, the T1 signal becomes brighter in adults. Thus, hypointense T1-weighted images of the clivus relative to the pons should be considered suspicious in adults. On T2-weighted sequences, clivus is usually isointense relative to the pons. Commonly, contrast with gadolinium does not show a considerable enhancement in healthy patients [5].

**Differential Diagnosis**

Clival lesions comprise tumoral and non-tumoral pathologies. Malignant neoplasms are more common than benign tumours in this location. Despite the benign behaviour of some entities, they can also be life threatening by compression of noble structures. Chordomas are the most frequent in this region and represent about 40% of all cases. Non-chordomatous lesions are rare and much diversified [8]. The main differential diagnoses are presented in the Table (1).

**Pituitary Adenomas**

Pituitary adenomas are the most common cause of sellar and...
Malign Neoplasms | Benign Neoplasms | Non-tumoral lesions
--- | --- | ---
Chordoma | Pituitary adenoma | Echordosis
Chondrosarcoma | Petroclival meningioma | Fibrous dysplasia
Metastasis | Osteoblastoma | Dermoid cyst
Multiple myeloma | Condroblastoma | Epidermoid cyst
Plasmocytoma | | Arachnoid cyst
Lymphoma | | Neurenteric cyst
Osteosarcoma | | Arachnoid herniation

Table 1: Differential diagnosis.

parasellar mass, comprising about 10 to 15% of all intracranial tumors. Despite histologically benign, they have the capacity to invade adjacent structures, such as: sphenoidal sinus, cavernous sinus, skull base or nasopharynx. The clivus invasion is rare and basi sphenoid is the most commonly affected portion [9]. Ectopic pituitary adenomas are extremely unusual and arise from residual cells along the migration tract of the anterior portion of the pituitary gland, during passage from Rathke’s pouch to the sellar turcica. Ectopic locations include the suprasellar region, sphenoid sinus, cavernous sinus and clivus. In these cases pituitary gland is commonly normal. However, purely ectopic clival pituitary adenomas are exceedingly rare. Pituitary adenomas classically present with bitemporal hemianopsia, however ectopic adenomas usually do not, unless they involve the optic chiasm. Headache is a common complain, but it is unspecific and cannot distinguish a pituitary adenoma from other lesions. Invasive and ectopic pituitary adenomas can manifest as a clival neoplasm in the imaging study and lead to misdiagnosis [10-12]. CT-scan typically reveals an enhancing soft-tissue mass that shows bone destruction. On T1-weighted MRI, pituitary adenomas demonstrate an isointense to slightly hypointense mass that enhances after administration of contrast (Figure 1). On T2-weighted images, they range from isointense to hyperintense, unless they contain intratumoral hemorrhage or cystic changes [13].

**Petroclival Meningioma**

Meningiomas are benign and slow-growing tumors that originate from the arachnoidal cells. The majority of them occur intracranially and they account for about 20% of all primary intracranial tumors in adults. Meningiomas are more frequent in women and their incidence increases with age. Intracranially meningiomas can be classified according place of origin and cellular type. The clivus is covered with dura mater, therefore it can be affected. Meningiomas at the center of the clivus proper are very uncommon. Petroclival meningiomas arise in the upper two thirds of the clivus at the petroclival junction, medial to the trigeminal nerve. They have a close relationship with the brainstem and large tumours can displace it or encase the basilar artery. The symptoms depend on the size of the tumor and they usually develop as a result of compression of surrounding structures. These patients commonly present with insidious onset of headache, seizures, unilateral hearing loss, facial sensory disturbances or trigeminal neuralgia [14,15]. The CT-scan reveals a hyperdense enhancing lesion and approximately a quarter of all cases demonstrate calcifications. CT-scan images may also be helpful in determining if the meningioma invades the bone, showing an intense hyperostotic bony response. T1-weighted MRI is generally isointense to slightly hypointense relative to the brain. Meningiomas vary from hypointense to hyperintense on T2-weighted images. Approximately 50% are isointense, 40% hyperintense and 10% hypointense relative to the gray matter. They reveal a strongly and homogeneous contrast enhancement, being the most sensitive method for detecting meningiomas. Angiography with vertebral and carotid studies is usually needed to elucidate the blood supply to the tumor and their relation with the major arterial branches [15, 16].

**Fibrous Dysplasia**

FD is a developmental disorder characterized by the progressive replacement of mature bone by fibrous tissue, caused by an abnormal proliferation and maturation of fibroblasts. This benign disease of unknown etiology results in the formation of woven and structurally weak bone. FD is more frequent in the first two decades of life and it may be present in the monostotic form (70%), affecting a single bone, or in the polyostotic form (30%), involving multiple bones. The polyostotic form can also occur as a part of McCune-Albright syndrome [17,18]. Craniofacial skeleton is involved in 30% of patients with monostotic disease and in 50% of patients with the polyostotic variant, affecting sphenoid, frontal, ethmoid, temporal and maxilla bones. FD rarely affects the clivus and when it occurs, does not seem to produce any functional limitation or aesthetic problem. However, clivus involvement may present headache, neck pain, craniocervical instability or hypoglossal nerve palsy [17,18].

Diagnosis of FD is based in the imaging findings and the high-resolution CT scan is the tool of choice. The characteristic appearance of the disease is ground-glass opacity, ballooning of the affected bone and thinning of the cortex. However, FD may present several patterns: ground glass, sclerotic, cystic or mixed pattern. The MRI appearance is less specific than CT. FD usually shows low signal intensity on T1-weighted images and variable signal intensity on T2-weighted images. Contrast enhancement is intermediate. This variation is determined by the disease activity, overall cellularity, extent of bone trabeculae, mineralization and cyst formation. Clival isolated lesions with low or intermediate signal intensity on T2-weighted MR imaging are rare. However, when FD presents high intensity on T2-weighted images, differential diagnosis is much more complex [19, 20].

**Cysts and Herniations**

Cysts are other important differential diagnoses in clival lesions. They can be divided in epidermoid, dermoid, neuroenteric and arachnoid cysts.

Epidermoid cysts are congenital lesions which result from inclusion of ectodermal elements during neural tube closure. They have a thin capsule of squamous epithelium and they can be smooth or lobulated. Internal components are often filled with desquamated epithelial keratin and cholesterol crystals. Epidermoid cysts are usually less sharply defined than arachnoid cysts and more heterogeneous. On MRL, the T1-weighted signal intensity tends to increase with the lipid content. The lesions are
Table 2: Imagological characteristics of different lesions.

<table>
<thead>
<tr>
<th>Histological Diagnosis</th>
<th>CT</th>
<th>T1-MRI</th>
<th>T2-MRI</th>
<th>Contrast Enhancement</th>
<th>Type of Enhancement</th>
</tr>
</thead>
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<tr>
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<td>Hypo/Isointense</td>
<td>Hyperintense</td>
<td>High</td>
<td>Heterogeneous</td>
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<tr>
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<td>Hypointense</td>
<td>Hyperintense</td>
<td>High</td>
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Figure 1: Coronal and sagittal T1-weighted images revealing a giant pituitary adenoma.

usually isointense relative to cerebrospinal fluid (CSF) on T2-weighted images. Epidermoid cysts present markedly restricted diffusion, different from arachnoid cysts [21].

Dermoid cysts are inclusion cysts composed of ectodermal elements such as hair, fluid, skin glands and rich in fat. Low attenuation values are suggestive of the diagnosis of dermoid cyst on CT. They also are hyperintense on T1-weighted images and hypointense on T2-weighted images, similar to fat [22].

Neurenteric cysts are endoderm derived lesions with ovoid/lobulated aspect. Neurenteric cysts of the clivus are unusual causes for clival masses. On CT-scan, they usually appear as a lytic lesion with intact cortex. The signal intensity depends of the protein content. They are isointense to slightly hyperintense compared with CSF on T1 W images, without contrast enhancement. Neurenteric cysts may show mild restriction on diffusion-weighted images [23].

Arachnoid cysts are avascular lesions containing CSF. Arachnoid cysts at the clivus are extremely rare. On the CT-scan, the margins of the lesion are well defined and sharp. They are unilocular, homogeneous, non enhancing and noncalcified and they present CSF density. On MRI, the signal is usually isointense on T1and T2-weighted images [24].

Intracranial hypertension can originate arachnoid herniations or lead to spontaneous CSF leaks. Theses nontraumatic osseous defects occur most frequently within the ethmoid and sphenoid sinus. In the clivus, the defects usually penetrate the posterior wall of the sphenoid sinus allowing for small arachnoid pits to herniate into the sinus [24].

**Chordoma**

Chordomas are slow-growing malignant tumors that arise from notocord remnants along the length of the neuraxis. These neoplasms have an incidence about 0.08 per 100,000 and they are more frequent in fourth and fifth decades of life. Skull base appears to be responsible for about 40% of all chordomas and the clivus is commonly involved location [25, 26]. Chordomas are locally aggressive and tend to recur after surgical resection,
however they rarely metastasize to distant sites. Patients with clival chordoma usually present with headache and diplopia, secondary to abducnt nerve palsy. Progressive compression and local invasion of the lesion may lead to other lower cranial nerve deficits [7, 26]. CT-scan typically shows an expansive soft-tissue mass arising from the intraosseous part of the clivus and causing extensive lytic bone destruction and showing intratumoral calcifications. Surrounding sclerosis is not common. There is heterogeneous enhancement following contrast administration. At MRI, chordomas are isointense to slightly hypointense on T1-weighted images and hyperintense on T2-weighted images, with areas of decreased signal intensity due to calcifications (Figure 2). Contrast attraction reveals a strong enhancement in both techniques, distinguish it from Echordosisphysaliphora (EP) [26-28].

Chondrosarcoma

Chondrosarcomas are malignant mesenchymal tumors composed of cartilage-producing cells. They can be developed de novo, resulted of a malignant transformation of a chondroma or can be associated with other osseous diseases. These tumors are more frequent in men in the fourth decade of life. Intracranial chondrosarcomas are more common in the skull base, arising from chondrocytes within cartilaginous remnants in petroclival and sphenoid-occipital synchondroses. They represent about 6\% of skull base neoplasms and they constitute a challenge, given their location. Despite they are overall slow growing, they manifest local aggressiveness. Skull base chondrosarcomas usually present with insidious onset headache and cranial nerve palsies such as hoarseness, dysphagia, diplopia, facial dysesthesia or hearing loss [29,30]. CT findings include enhancing soft-tissue mass with a calcified matrix, bone erosion and destruction and a sharp zone of transition to normal tissue. CT-scan is useful for detecting subtle calcifications in the matrix when the diagnosis is in doubt. On MRI, lesions are hypointense on T1-weighted images and hyperintense on T2-weighted images, relative to the brain (Figure 3). The contrast enhancement is very strong. Neoplasms commonly exhibit heterogeneous internal areas of decreased signal, corresponding to internal calcifications [31].

Metastasis

Metastatic lesions to skull base are infrequent. Clivus metastases commonly arise from prostate, lung breast and head and neck cancer. Hematogenous spread of cancer cells to the clivus can occur via two routes: a) through the Batson plexus into the internal vertebral venous plexus system, which communicates superiorly with the basilar plexus at the clivus; b) or via the inferior ophthalmic vein, which drain into the cavernous sinus and communicates with the inferior petrosal sinus and basilar plexus. Cranial neuropathies are the main expression of skull base metastases and abducens palsy can occur in the clivus invasion [31,32].

CT-scan of the clivus metastases reveals areas of lytic bone destruction, except prostatic cancer, which is usually osteoblastic. T1-weighted MRI shows replacement of normal bone marrow by material of decreased signal intensity. On T2-weighted images, metastases are generally hyperintense and the addition of contrast material usually reveals enhancement.

However, imaging alone does not allow differentiation between primary and metastatic cancer. FDG-PET/CT has an important role in identifying an unknown primary or progression of a prior tumor [33,34].

Lymphoma, Plasmacytoma and Multiple Myeloma

Primary bone lymphoma is a rare malignancy, accounting for less than 5\% of all primary bone tumors. Femur and pelvis are the most common affected locations, although the clivus can also be involved. CT-scan reveals a lytic or blastic-sclerotic pattern with cortical breakthrough and peristomal reaction. MRI reveals areas of low signal intensity within the marrow on T1-weighted images. These areas generally appear bright on T2-weighted sequences, as well as peri-tumoral edema and reactive marrow changes [35].

Plasmacytoma and multiple myeloma are clonal neoplasms of terminally differentiated plasma cells. Plasmacytoma is a localized collection of plasma tumour cells and usually presents as a solitary lytic lesion. Multiple myeloma is a multifocal and destructive tumour involving the axial skeleton with features of osteolytic lesions, pathological fractures, hypercalcaemia, bone pain and serum monoclonal protein levels. The most commonly affected bones are the vertebrae, ribs, skull and pelvis. In the skull base, plasmacytomas can affect clivus as a solitary lesion of challenging diagnosis [36]. On CT-scan, these neoplasms appear as a punched-out lytic lesion without sclerotic rims and an expansive soft-tissue mass. On MRI, T1-weighted images generally show a focal hypointense area and T2-weighted images and gadolinium-enhanced images present a hiperintense focal lesion relative to the hypointense marrow background. Scintigraphy has a limited interest because mainly detect osteoblastic response of the skeleton and plasma cells tumours are osteolytic lesions, underestimating the extent of the disease [36]. The FDG-PET/CT shows focal uptake in the metabolically active lesion and allows the detection of other involved places in multiple myeloma. FDG-PET/CT also reveals different patterns of Fluorodeoxyglucose uptake, according the activity of the tumour. Plasmacytomas show a mild-intense signal, helping in the differential diagnoses of solitary clivus neoplasms [37].

Osteosarcoma

Osteosarcomas are the most common primary bone malignancies. They are highly aggressive lesions that comprise osteoid-producing spindle cells. Osteosarcomas usually occur in the second decade of life and may be associated with Paget disease, Fibrous dysplasia or prior radiation. Predominant locations are the metaphysis of long bones but skull base can also be affected. CT-scan reveals a destructive mass involving the clivus with multiple ossifications and slight contrast enhancement. On MRI, T1-weighted images show an isointense mass with expansive appearance and T2-weighted images are slightly hyperintense [38].

Chondroblastoma

Chondroblastomas are benign primary bone tumors that originate from chondroblasts. They typically develop in the epiphyses of long bones, however skull base including clivus can also be affected. Chondroblastomas usually present in the second
In the decade of life, with a male predominance. When they implicate the clivus, the patient can refer diplopia and headache. CT-scan reveals an osteolytic lesion with areas of punctate calcifications and sclerotic margins. On the MRI, they usually have low-to-intermediate intensity on T1 and low-to-high intensity on T2 with variable contrast enhancement following gadolinium [39].

**Osteoblastoma**

Osteoblastomas are uncommon primary bone neoplasms with slow-growing and benign behavior. Clivus involvement is uncommon, but it can occur. CT-scan presents a well-circumscribed osteolytic and multiloculated mass, mottled by calcifications and with an intact cortex. T1-weighted images are hypointense and T2-weighted images are hyperintense with contrast enhancement [40].

**Ecchordosis Physaliphora**

Ecchordosis Physaliphora (EP) is a tumor-like congenital malformation which arises from ectopic notochordal remnants, located along the midline craniospinal axis, from the clivus to the sacrococcygeal region. Although these remnants usually have an intraosseous location, they occasionally perforate through the dorsal wall of the clivus. Intracranial EP is typically found in the prepontine cistern and attached to the dorsal wall of the clivus by a small pedicle. EP is habitually asymptomatic and found incidentally, however tumor expansion and compression of surrounding structures were reported in some cases [41,42].

On CT-scan, these lesions reveal focal sclerosis without bone destruction, different from chordomas. EP is well-circumscribed and homogeneously hypointense on T1-weighted images and hyperintense on T2-weighted images, without enhancement after gadolinium addition. This lack of enhancement is also useful to distinguish it from chordomas and other malignant tumours [42,43].

**Differential Diagnosis**

Clivus is a sloping structure that forms the anterior portion of the posterior cranial fossa, in the center of the skull base. Due to its strategic location and its embryologic origin, some disorders typically affect the clivus. Clival lesions are rare and they represent a wide range of pathologies, including non-tumoral lesions, benign and malignant neoplasms. Chordomas account for about 40% of all entities, although it is often difficult to distinguish them from non-chordomatous lesions [2, 8].

Clinical symptoms and signs of clival disease are nonspecific or absent. Headache and cranial neuropathies, especially abducent nerve palsy, are the most common complaints. Furthermore, these lesions often present as an incidental imaging finding [3].

Imaging study constitutes a useful tool in differential diagnosis. CT-scan is the best exam for evaluating cortical bone and for demonstrating the detailed anatomy of the cortical margins of the clivus. However, CT has less sensitivity in evaluating bone marrow space involvement until significant marrow infiltration and subsequent trabecular bone loss has occurred. CT-scan is important in primary bone lesions, distinguishing bone tumors from fibrous dysplasia or other bone disease. One the

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**Figure 2** Intra-operative images of a clival chordoma on neuronavigation system.
other hand, MRI is more sensitive in assessing bone marrow signal abnormalities in the skull base and in discriminating soft tissue. In adults, hypointense T1-weighted images of the clivus relative to the pons should be considered suspicious. On T2-weighted sequences, clivus is usually isointense relative to the pons [5]. Hyperintensity on T2-weighted images and strong contrast enhancement with gadolinium suggest chordoma and chondrosarcoma (Table 2). MRI also gives additional information about dural infiltration and the intracerebral origin of the tumor [5, 31]. Despite imaging study, a final diagnosis cannot be achieved without histologic analyses because, sometimes, lesions are radiologically difficult to specify. Thus, an accurate diagnosis remains a challenge and needs a close cooperation between surgeon, radiologist, and pathologist in a multidisciplinary team work.

Chordomas and chondrosarcomas are difficult to distinguish because both of them present hypointense signal on T1-weighted images, hyperintense signal on T2-weighted images and contrast enhancement with gadolinium [26, 29]. Thus, definitive diagnosis is based on the immunohistochemical study. Chordoma reacts with cytokeratin and epithelial membrane antigen (EMA), whereas chondrosarcoma is negative for cytokeratin and EMA [44]. Clival metastases also are an important differential diagnosis. They can occur in the context of a known history of a distant malignancy or can be the presenting symptom. Imaging alone does not allow differentiation between primary and metastatic cancer. FDG-PET/CT may help in identifying an unknown primary or progression of a prior tumor [33,34]. FDG-PET/CT is also useful on plasma cells neoplasms because it reveals different patterns of Fluoro deoxyglucose uptake and allows the detection of other involved places in multiple myeloma. Plasmacytomas show a mild-intense uptake, different from other clival lesions [37]. The histologic and immunohistochemistry similarity reinforce the possibility that chordomas may arise from EP. On MRI study, chordomas commonly enhance markedly with gadolinium, although EP does not show any enhancement with contrast addition [42,43]. Non-tumoral lesions as FD or cysts also need to be considered in the differential diagnosis. Given their very slow growth potential, asymptomatic FD of the clivus and cysts can be usually treated with clinical observation [36].

CONCLUSION

Clival disorders comprise a large range of entities with different characteristics and behaviors. The knowledge of these pathologies is crucial to establish a correct workup and achieved the right diagnosis. Imaging study allows an evaluation of the tumor extent, a reliable distinction between benign and malignant processes and helps in the suspicious diagnosis. Despite imaging is an important supporting element, a final diagnosis often cannot be found without histologic biopsy. Thus, it is necessary a multidisciplinary team with close cooperation between surgeon, radiologist and pathologist to achieved the correct diagnosis.

REFERENCES


