

Case Report

Complicated Pansinusitis Masquerading Orbital Apex Syndrome

M. Silva*, J.C. Ribeiro, J. Romão and A. Paiva

Department of Otolaryngology and Head and Neck Surgery, Universitário de Coimbra, Portugal

*Corresponding author

Miguel Silva, Department of Otolaryngology and Head and Neck Surgery, Universitário de Coimbra, R. Fonseca Pinto, 3000-075 Coimbra, Portugal, Tel: 239-400-400; Email: stoj_@hotmail.com

Submitted: 12 June 2015

Accepted: 03 July 2015

Published: 06 July 2015

Copyright

© 2015 Silva et al.

OPEN ACCESS

Keywords

- Intraorbital plasmacytoma
- Orbital tumor
- Orbital apex syndrome

Abstract

Orbital Apex Syndrome is a very rare clinical diagnosis presented to ENT surgeons. Intraorbital extramedullary plasmacytoma is extremely rare and there are a few cases reports in the literature. Very unusually it develops a secondary leukemic transformation to a plasma cell leukemia which is a very rare and aggressive type of acute leukemia (in our tertiary hospital, Hematology Department had previously 3 cases in the last fifteen years).

Orbital apex syndrome is a rare but potentially fatal situation rarely seen initially by ENT surgeons. We report an even rarer case of an intraorbital extramedullary plasmacytoma that caused orbital apex syndrome, initially masqueraded by a complicated pansinusitis.

An immunosuppressed 78-year-old female presented with a progressive left side reduced best corrected visual acuity, pansinusitis and orbital involvement, imaging showed opacification of all paranasal sinuses, cavernous sinus involvement and left intraorbital mass lesion. After an initial improvement with antibiotics, sudden clinical deterioration occurred. Biopsy of the intraorbital mass showed a monoclonal lymphoid neoplasm and laboratory findings led us to the diagnosis of plasma cell leukemia.

We advocate for a multidisciplinary approach ab initio in all cases of complicated rhinosinusitis to minimize or prevent deterioration of vision and optimize clinical outcomes.

INTRODUCTION

The orbital apex is the most posterior part of the orbit, positioned at the craniofacial junction. Infectious, inflammatory, neoplastic, traumatic and iatrogenic conditions in this region may cause an Orbital Apex Syndrome (visual loss from optic neuropathy, ophthalmoplegia involving multiple cranial nerves and eyelid ptosis) [1,2]. It is a rare syndrome, more often presenting initially to the ophthalmologist. Because it is a potentially fatal condition frequently causing severe morbidities, ENT surgeons must be aware of it nevertheless it seldomly presents first to them [2].

We report an intraorbital extramedullary plasmacytoma that came out as an orbital apex syndrome, initially masqueraded by a complicated pansinusitis.

There are only few cases reports about intraorbital plasmacytoma with secondary plasma cell leukemia. Even rarer is the fact that it presented initially by orbital apex syndrome and pansinusitis.

CASE REPORT

A 78-year-old female with a history of chronic immunosuppression with cyclosporine (renal transplanted 11 years ago), bilateral open-angle glaucoma diagnosed 8 years ago, history of coronary bypass surgery 7 years ago, new onset diabetes mellitus type II presented to ENT emergency room with bilateral nasal obstruction, mucopurulent rhinorrhea and facial pain with more than 8 days of evolution. Relevant clinical findings included bilateral anterior and posterior mucopurulent rhinorrhea and tenderness on facial percussion. Best corrected visual acuity (BCVA) for each eye was 1/10. She was discharged with a diagnosis of acute rhinosinusitis and treated with amoxicillin 875mg and clavulanate 125mg twice daily, paracetamol for pain control and nasal lavage. One week after treatment, the patient showed up in the ENT emergency room with worsen mucopurulent rhinorrhea, nasal edema and initially with sudden decreased left visual acuity, later with superior palpebral ptosis and complete ophthalmoplegia on the left side. Ophthalmologist was consulted, BCVA was 3/60, no alteration on the right eye

was detected, intraocular pressure remained in the normal range and there was no other relevant clinical finding. There were no cervical or axillary lymphadenopathy, no abdominal mass.

Laboratory studies showed hemoglobin level of 11mg/dl, leucocytosis ($13 \times 10^9/L$) with neutrophilia, normal hepatic function, creatinine level of 1.34mg/dL, C-reactive protein level of 17.5 and erythrocyte sedimentation rate of 30mm/h. Paranasal and orbital CT- scan showed a pansinusitis with metal density spots and a diffuse left intraorbital apex opacification with a mass-like lesion mixed up with the superior rectus muscle (Figure 1,2). She was admitted to the ENT ward. Initially, the clinical diagnosis was orbital apex syndrome caused by fungal rhinosinusitis. Differential diagnosis included intra-orbital tumor, Tolosa-Hunt syndrome and cavernous sinus thrombosis. Therefore, systemic broad spectrum antibiotics (ceftriaxone 2g/day), amphotericin B (3mg/kg ivqday) and systemic corticosteroids (125mg methylprednisolone twice daily) were immediately started. At beginning, patient showed a slight clinical improvement. No improvement was noted in the affected left eye. Cranio-encephalic and orbital MRI showed bilateral decreased cavernous sinus flow and a left intraorbital diffuse soft tissue mass involving the superior rectus muscle (Figure 2,3). *Staphylococcus aureus* was cultured from middle meatus rhinorrhea and there was no fungal growth. Viral serology and auto-immunity markers were all negative. After another week without further improvement, the intraorbital mass was biopsied through endoscopic endonasal approach and showed a monoclonal lymphoid neoplasm with an inconclusive phenotype. After three weeks of admission,



Figure 1 Orbital CT-Scan showing a mass-like lesion in the left orbital apex and diffuse heterogeneous opacification of the paranasal sinuses.



Figure 2 Orbital CT-Scan showing a mass-like lesion in the left orbital apex.

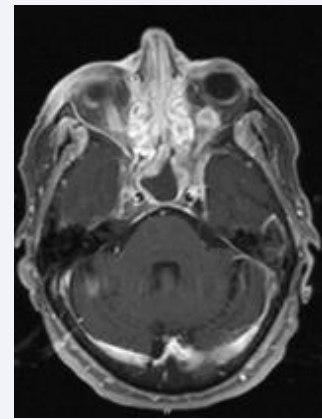


Figure 3 Orbital MRI confirmed a tumor in the left orbital apex.

patient suddenly developed a progressive elevation of leukocytes counts to $60.5 \times 10^9/L$ and peripheral smears detected an abundant amount of blast cells. Flow cytometry, cytogenetic and immunophenotype analysis led to the diagnosis of plasma cell leukemia, most probably by secondary leukemic transformation of left intraorbital plasmacytoma.

Patient's clinical condition had deteriorated rapidly with multi organ failure and died 30 days after initial clinical manifestation.

DISCUSSION

Acute rhinosinusitis is a common condition treated by ENT surgeons. Major complications like cavernous sinus thrombosis are unusual nowadays due to an easy access to antibiotic treatment.

This patient showed initially a clinical condition that resembled pansinusitis complicated with cavernous sinus thrombosis. A thorough investigation led us to the final diagnosis of an intraorbital plasmacytoma with plasma cell leukemic transformation.

Orbital apex syndrome has been described as a condition hallmarked by optic nerve, oculomotor nerve, trochlear nerve, abducens nerve and ophthalmic branch of trigeminal nerve paralysis [1]. The common causes of this condition are neoplasm, inflammatory and infectious diseases [2]. Fungal origin should be suspected in immunocompromised patients [3,4]. The cavernous sinus syndrome may include the features of an orbital apex syndrome with added involvement of the maxillary branch of the trigeminal nerve and oculosympathetic fibers. Cavernous sinus lesions are more commonly bilateral [2]. Because of patient's history of immunosuppression with cyclosporine and new-onset diabetes associated with distinct CT-scan images, initial diagnosis was fungal pansinusitis. However, there was no improvement with aggressive medical treatment. Endoscopic endonasal biopsy of the mass lesion in the orbital apex, rapid progression of the disease and elevation of leukocytes count led us to a neoplastic etiology. There are several types of plasma cell neoplasm: multiple myeloma, plasma cell leukemia, intramedullary and extramedullary plasmacytoma. The last one is an unusual condition and represents 3% of all plasma cell neoplasm, most often occur in the paranasal sinus, nasopharynx and larynx [5].

In literature, there are few cases of intraorbital extramedullary plasmacytomas and very unusually it developed a secondary leukemic transformation to plasma cell leukemia, a very rare condition and one of the most aggressive neoplasms [6]. Three days after the diagnosis of plasma cells leukemia, her clinical condition deteriorated and she died.

With such a highly aggressive nature of the tumor, unequivocal initial diagnosis usually arrives too late. We recommend involving ophthalmologists early in a multidisciplinary approach to patients with complicated sinusitis with atypical signs and symptoms of the eyes because a detailed orbital evaluation may prevent missing important and progressive diagnoses. We also recommend performing an MRI in all patients with equivocal complicated sinusitis or unilateral ophthalmoplegia that have not improved after 72h of treatment. A more aggressive approach shall be performed in this kind of patients because it is potentially fatal. Management is directed at the underlying cause and may be guided by surgical biopsy.

CONCLUSION

Prompt diagnosis and treatment in cases of orbital apex syndrome, especially in immunocompromised patients, is important to prevent visual and life-threatening complications.

Detailed orbital evaluation should be considered in all patients who present any atypical signs and symptoms of the eyes, to prevent missing important and progressive diagnoses.

REFERENCES

1. Bone I, Hadley DM. Syndromes of the orbital fissure, cavernous sinus, cerebello- pontine angle, and skull base. *J Neurol Neurosurg Psychiatry*. 2005; 76 Suppl 3: iii29-29iii38.
2. Yeh S, Foroozan R. Orbital apex syndrome. *Curr Opin Ophthalmol*. 2004; 15: 490-498.
3. Rao SP, Kumar KR, Rokade VR, Khanna V, Pal C. Orbital Apex Syndrome due to mucormycosis caused by *Rhizopus microsporum*. *Indian J Otolaryngol Head Neck Surg*. 2006; 58: 84-87.
4. SH Lee, SM Hong, HM Lee. Orbital Apex Syndrome Related to Isolated Sphenoid Fungal Sinusitis. *J Rhinol*. 2013; 20: 123-126.
5. Kelly SP, Lloyd IC, Anderson H, Joyce PW, Pace-Balzan A. Solitary extramedullary plasmacytoma of the maxillary antrum and orbit presenting as acute bacterial orbital cellulitis. *Br J Ophthalmol*. 1991; 75: 438-439.
6. Morley AM, Verity DH, Meligonis G, Rose GE. Orbital plasmablastic lymphoma--comparison of a newly reported entity with diffuse large B-cell lymphoma of the orbit. *Orbit*. 2009; 28: 425-429.

Cite this article

Silva M, Ribeiro JC, Romão J, Paiva A (2015) Complicated Pansinusitis Masquerading Orbital Apex Syndrome. *Ann Otolaryngol Rhinol* 2(6): 1047.