Case Report

Retroperitoneal Ancient Schwannoma: A Rare Case Report

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Abstract

Retroperitoneal ancient schwannoma is a quite uncommon tumor of the peripheral nerve sheaths. Despite its benign course, it may also display malignant behavior. Herein, we presented a retroperitoneal ancient schwannoma case together with clinical, radiological and pathological characteristics of the disease, which had been detected incidentally via ultrasonography in a 24-year-old patient with epigastric pain.

ABBREVIATIONS

MRI: Magnetic Resonance Imaging; SMA: Smooth Muscle Action; CT: Computed Tomography

INTRODUCTION

Ancient schwannoma is a rarely encountered variant of retroperitoneal schwannoma. There is limited number of publications on this subject in the literature [1,2]. Schwannoma, otherwise known as neurilemmoma, is a benign soft tissue tumor arising from Schwann cells of the peripheral nerve sheaths. Whilst schwannomas are usually seen on the flexor surface of the head, neck and extremities, they are quite rarely seen in the retroperitoneal region [3]. Of all schwannomas, 0.3-3.2% are found in the retroperitoneal region [4]. In general, they are seen between the ages of 40-60 years and in females more frequently than males [5]. It can either be sporadic or may be seen as the component of genetic syndromes such as neurofibromatosis and schwannomatosis [6]. As in the present case, tumors of peripheral nerves should be considered in the differential diagnosis of the masses of this region and surgical excision should be prioritized considering that biopsy is not a procedure yielding definite outcome.

CASE PRESENTATION

The 24-year-old male patient, who presented with epigastric discomfort, underwent magnetic resonance imaging due to retroperitoneal cystic mass seen on ultrasonographic examination. MRI demonstrated approximately 8 cm cystic mass in front of vertebral column adjacent to the right kidney [Figure 1]. Urogenital examination of the patient was unremarkable. Alpha fetoprotein (AFP), beta human chorionic gonadotropin (beta-HCG) and lactate dehydrogenase (LDH) were within the normal ranges. Scrotal color Doppler ultrasonographic and thoracic tomographic examinations were considered normal. The mass was surgically excised via right-side chevron incision. The mass was 8x6 cm in size and had round shape with smooth surface [Figure 2]. Histopathological examination of the mass revealed degenerated ancient schwannoma [Figure 3 and 4]

DISCUSSION

Schwannomas are slow-growing, well circumscribed, non-aggressive, encapsulated tumors with smooth surface and are generally diagnosed incidentally [7]. They are usually asymptomatic, but may be symptomatic when they grow larger. The symptoms are most frequently nonspecific such as abdominal pain and discomfort [8]. More rarely, varicose veins, headache, hypertension, hematuria, or renal colic may be seen [7].

Ancient schwannoma is a rare morphological variant. It was first defined in 1951 by Ackerman and Taylor [9]. This definition indicates histological degenerative character, which develops with the growth and aging of the tumor. Nuclear atypia of the tumor makes it difficult to be discriminated from malignant tumors [8]. Central cystic degeneration, perivascular hyalinization, hemorrhage, necrosis, pleomorphic hyperchromatic nuclei, and calcification seen in these larger tumors indicate that schwannoma has existed for a long time [10]. Histologically, presence of low mitotic activity and Kern-Loch phenomenon, which are associated with atypical nuclei of the ancient schwannomas, helps in the diagnosis [11]. In immunohistochemical studies, the tumor shows positive staining with S-100, vimentine and neuron-specific enolase but negative staining with smooth muscle action [SMA] and CD 117 [12].
the schwannomas, cellular and fusiform cells are microscopically seen in the Antoni A areas; however, Antoni B areas are poor in cells and contain plenty of myxoid matrix [1]. The mass in the present case had cystic degeneration, hemorrhage and necrosis and showed immunohistochemical positive staining with S-100.

The majority of the schwannomas are benign. Malignant retroperitoneal schwannomas are rare tumors with poor prognosis and usually exist as a component of Von Recklinghausen syndrome [7,13]. These tumors may show local recurrence or distant metastasis [14]. There is no histopathological standardized diagnostic criterion for malignant schwannomas. However, asymmetric, conic, fusiform cells may contain massive fascicles like a marble, and there may be increased mitosis, pleomorphism, and blood vessel infiltration [15].

CT and MRI are the methods used most frequently although they are preoperatively nonspecific. In addition, fine needle aspiration biopsy can be performed easily and safely [16]. However, there are reports suggesting that biopsy has poor diagnostic value and may lead to misdiagnosis; moreover, it may cause bleeding, infection or neoplastic spread [17]. As there is no method that could preoperatively discriminate malignant from benign tumors, surgical resection of tumor is the most appropriate approach. Frozen could be performed during surgery if it is necessary to determine the negative surgical margin accurately. The rate of local recurrence after resection is 16-56% for malignant tumors [8]. Radiofrequency ablation has been suggested as an option in the patients that are not suitable for surgical resection as it is minimally invasive, effective and repeatable [18]. In the present case, the mass was resected as en block, and frozen was not required to determine the negative margin because no mass-related tissue has been left.

Scanning methods and needle biopsy are inadequate for making diagnosis, and definite diagnosis is made by surgical resection and histopathological examination of the mass. Although schwannomas have generally good prognosis, the
patients should be cautiously followed for local recurrence. Histopathological examination report of the present case revealed benign degenerated ancient schwannoma and no relapse was encountered over the course of one-year follow-up period.

REFERENCES


