· Case report ·

Orbital ganglioneuroma: a case report

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Abstract

• We present a very rare case of orbital ganglioneuroma. A 13-year-old man presented with more than 5 years' history of exophthalmos of right eye. Computed tomography (CT) scan disclosed a tumor in the right orbit. The patient received surgical treatment. Pathological examination revealed a ganglioneuroma. The patient was followed up for 12 months without recurrence.
• KEYWORDS: orbit; ganglioneuroma
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INTRODUCTION

Ganglioneuroma is an uncommon benign neurogenic tumor which is derived from the embryologic neural crest[1]. It can arise anywhere. Here we present a very rare case of a 13-year-old man with an orbital ganglioneuroma. As we know this is the first report of ganglioneuromas in the orbit.

CASE REPORT

A 13-year-old man presented with more than 5 years' history of exophthalmos of right eye. In the year of eight, the patient was discovered with a right-sided exophthalmos without vision damage, pain and redness (Figure 1). Systemic examinations were normal. Eye examination showed the vision was 20/20 of both eyes. The exophthalmometer showed the distance from the cornea to the orbital rim of the right eye and the left eye was 18mm and 13mm, respectively. Compared with the left eye, the right eye was displaced upwards. The eye movement was normal. Bilateral anterior segments and intraocular pressures were normal. Funduscopy of both eyes also showed normal. Computed tomography (CT) scan revealed a mass in the region of the right inferotemporal orbit and pushed the eyeball upwards, the mass had clear border with even density (Figure 2). His routine blood, urine, liver and kidney function tests were normal. Chest X-ray was normal. The tumor was removed completely through an inferior orbitotomy approach. Postoperative ophthalmological examination showed visual acuity maintained 20/20 in both eyes. The eye movement was normal without eye displacement (Figure 3). The patient was followed up for 12 months without recurrence.

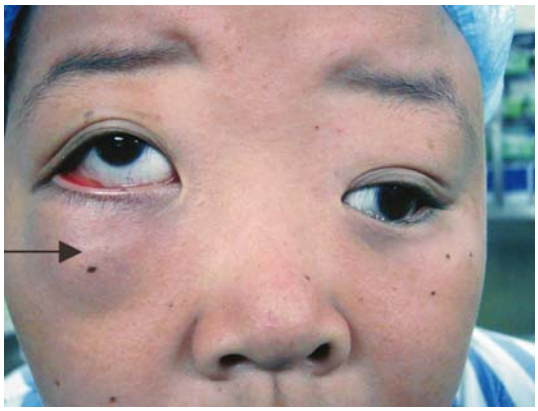

Figure 1 Preoperative extraocular photograph showed the mass located in the inferior right orbit (arrow), and pushed the eyeball upwards.

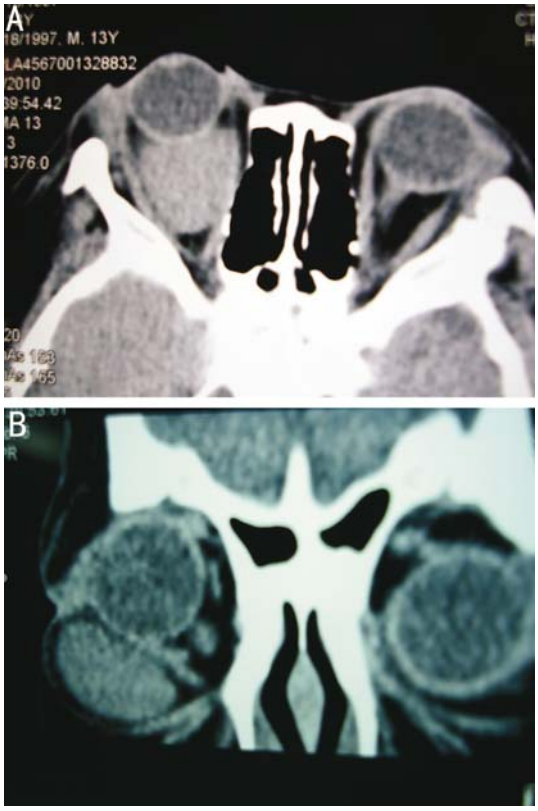

Figure 2 A: Axial CT scan revealed a well circumscribed solid mass of the right orbit; B: Coronal CT scan showed the mass located in inferotemporal aspect of the right orbit.

Histopathologic and Immunohistochemical Examination

Pathology: Grossly, the ganglioneuroma was well circumscribed with a fibrous capsule and the mass color was pink (Figure 4). Histologically, the tumor was consisted of longitudinal and transversal bundles of schwann cells that arranged in an irregular

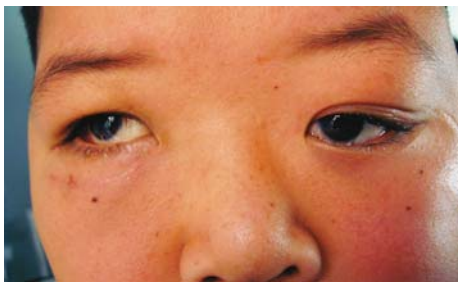

Figure 3　The right eye was almost in normal location one week after operation.

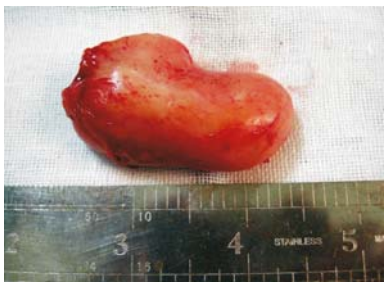

Figure 4　Gross sample shows intact fibrous capsule with pink color.

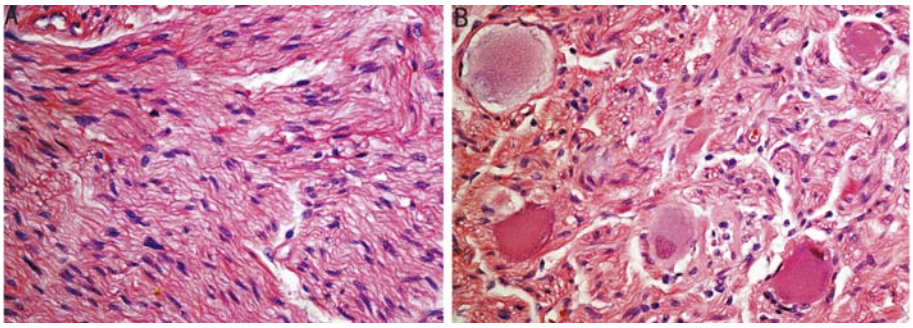

Figure 5　A:The background stroma was composed predominantly of schwann cells which presented spindle shape with bundle arrangement (HE ×400); B: The pink ganglion cells with one to three nuclei scatter the stroma(HE ×400).

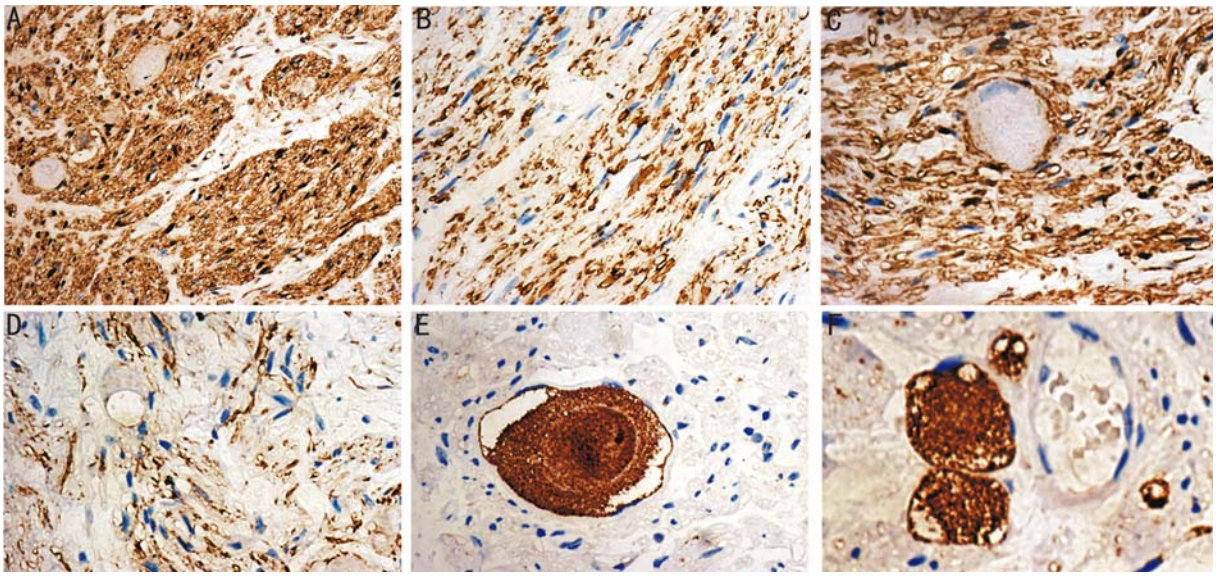

Figure 6　Immunohistochemical examination revealed fibers(+) ganglion cells(-) of S-100 in A, NF in B, Vimentin in C, GFAP in D, and ganglion cells(+) fibers(-) of NSE in E, Syn in F.

fashion in the background. There were some relatively mature ganglion cells which scattered throughout the stroma. The ganglion cells may occur in a big solitary fashion, or in small clusters. Their abundant cytoplasm was red pink and had one to three nuclei. Sometimes pigment was found in the cytoplasm (Figure 5). Immunohistochemically, it revealed that schwann cells were stained by S-100, neurofilament (NF), glial fibrillary acidic protein (GFAP) and vimentin, the ganglion cells were positive in neuron-specific enolase (NSE) and synaptophysin(Syn) (Figure 6).

DISCUSSION

Ganglioneuromas are tumors originating from neural crest cells. They often present in adolescents and young adults[2,3]. They usually occur in relation to tissues that originate from the neural crest and the paraspinal sympathetic chain ganglia. The most common locations of ganglioneuromas are the posterior mediastinum and retroperitoneum [4,5]. It is very rare present in orbit, to our knowledge, this is the first report of orbital ganglioneuroma. It probably arose from the small orbital sympathetic nerve terminal.

Ganglioneuromas can produce varying symptoms in association with their location and resultant mass effect. It can lead to painful, headache, paresthesia, hyperhidrosis, bowel or bladder dysfunction, concentration difficulties and memory disturbances[6-11]. Depending on the location of the tumor, the patient of our case with orbital ganglioneuroma presented with exophthalmos, eyeball displacement. The vision and the eye movement were normal. Histological examination shows

advanced neuronal differentiation with neurocytes and ganglion cells. Immunohistochemistry typically shows expression of NSE and Syn for ganglion cells [6]. Our case was consistent with it. Surgical resection is the most reasonable approach to therapy [12]. The patient received a complete excision of tumor and the prognosis is good, no recurrences have been described so far.

This case shows that ganglioneuroma should be added to the differential diagnosis of tumors that can occur in the orbit, including schwannoglioma, meningioma, neuroglioma, cavernous hemangioma, lymphomas, and metastatic tumors, *et al*.

CONCLUSION

Orbital ganglioneuroma is very rare. This case shows that ganglioneuroma should be added to the differential diagnosis of orbital tumors.

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眼眶神经节细胞瘤1例

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摘要

报道1例罕见的眼眶神经节细胞瘤:1例13岁男性右眼突出5a^{+},CT检查示右眼眶下方包块,通过手术完整切除包块,病理检查证实为眼眶神经节细胞瘤,患者随访12mo无复发。

关键词:眼眶;神经节细胞瘤