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Case Report

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SUPRASELLAR SCHWANNOMA MASQUERADING AS CRANIOPHARYNGIOMA: AN EXTREME RARITY

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INTRODUCTION

Intracranial schwannomas account for 8-10% of all primary brain neoplasms.[1] They are most frequently located in the cerebello-pontine angle and originate from the vestibular component of the VIII cranial nerve. Although, sensory nerves are the preferred sites of development, motor and autonomic nerves may also be involved.[2] However, intracranial schwannomas not related to cranial nerves are very rare and especially intra-suprasellar location of schwannomas has been reported very infrequently.[3-6] We, here, report an unusual case of an intra-suprasellar schwannoma that was initially thought to be a craniopharyngioma both clinically and radiologically but final histopathological diagnosis was a primary intrasellar schwannoma.

CASE REPORT

A 4 year-old male child was brought to neurology outpatient with complaints of headache and projectile vomiting for 2 months. There was no history of fever and trauma. General physical examination was within normal limits. Neurological examination revealed bitemporal hemianopia with poor visual acuity. The routine endocrine testing showed normal pituitary hormonal function with serum prolactin levels of 3.46 ng/ml and normal thyroid function tests. Magnetic resonance imaging revealed a bulky solid-cystic intra-suprasellar mass measuring 35x32x38 mm in size, which was hyperintense with brain parenchyma on T1-weighted imagesand iso-hyperintense on T2-weighted images (Figure 1). The lesion was causing mass effect on the optic chiasma and was extending into the third ventricle causing moderate dilatation of bilateral third ventricles. Posteriorly mass was seen extending into the interpeduncular and prepontine cisterns, with broad area of contact with basilar artery. A provisional diagnosis of neoplastic lesion likely to be acraniopharyngioma was suggested. Frontotemporal craniotomy was done and orbital roof was removed upto orbital apex. The dura was opened with its base towards temporal side. Cerebrospinal fluid was drained by ventriculoperitoneal shunting. Bilateral optic nerves were thickened with tumor bulging into interoptic space. So, an intraoperative diagnosis of optico-chiasmatic glioma was rendered. The mass was yellowish, elastic-in-consistency and hard and it was partially removed by means of curettage and suction. We received multiple gray brown soft tissue pieces measuring altogether 1.0 x 0.9 x 0.7 cm.

The histopathologic examination showed benign neurogenic tumor with reactive gliosis at the periphery, compatible with features of schwannoma. Light microscopically, the lesion was mainly composed of spindle cells arranged in small whorls and short fascicles. The individual cells showed elongated and wavy cytoplasm with indistinct cell borders. The nuclei were oval with fine to hyperchromatic chromatin and indistinct small nucleoli. Among the spindle cells, slit like capillaries were scattered. Focal microcystic changes, infiltration of inflammatory cells and extravasated red blood cells were also found. Neither mitosis nor necrosis was found. Under the light microscope, there were perivascular hyalinization orAntoni A, B including Verocay bodies (Figure 2). Immunohistochemically, there was high and diffuse positivity for the S- 100 protein and glial fibrillary acidic protein (GFAP, Figure 3).

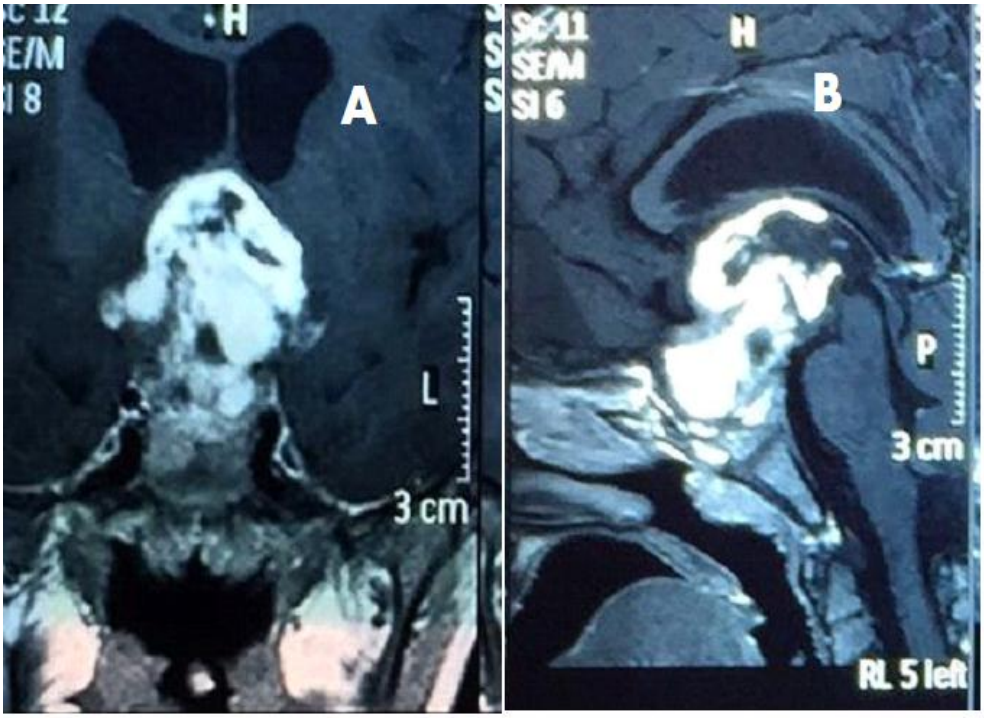

FIGURE 1: MAGNETIC RESONANCE IMAGING SHOWING INTRA- SUPRASELLAR HYPERINTENSE MASS LESION WITH DILATATION OF THIRD VENTRICLE.

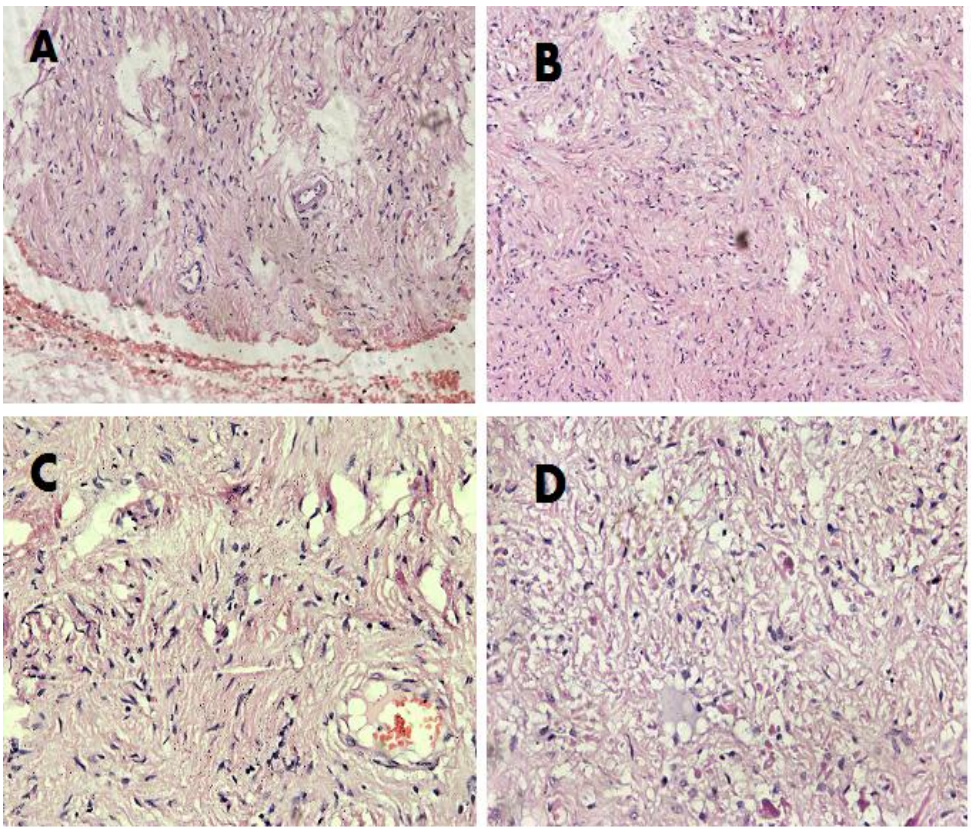

FIGURE 2: PHOTOMICROGRAPH SHOWING ENCAPSULATED BENIGN NEUROGENIC TUMOR (A) AT 40 X, (B) 200X, (C) AND (D) 400X.

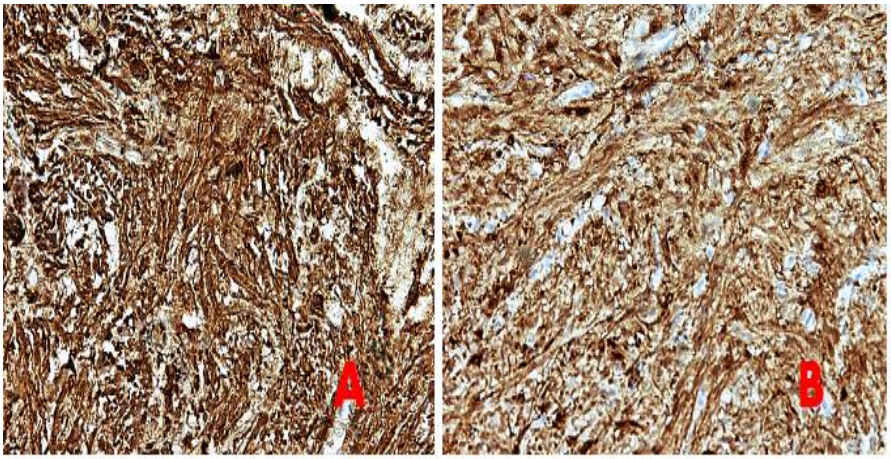

FIGURE 3: IMMUNOHISTOCHEMISTRY SHOWING GFAP (A) AND S100 (B) DIFFUSE POSITIVITY. (200X)

DISCUSSION

The space occupying lesions of the sellaturcica include pituitary adenomas, meningiomas, craniopharyngiomas, chordomas, germinomas and non-neoplastic cysts of inflammatory conditions and so on.[7] However, schwannoma is not usually included, because the occurrence of schwannoma in the sellar or suprasellar region is extremely rare and it can mimic a non-functioning pituitary adenoma.[8] Only twelve cases of intrasellar schwannomas have been reported in medical literature till date, all of which presented as suprasellar extension similar to our case.[3-6] Schwannomas account for about 8% of all primary intracranial neoplasms and are relatively frequent intracranial tumors. They usually develop from sensory nerves, most often the auditory nerve and less commonly from the trigeminal nerve, although they have been shown to involve all other cranial nerves except the optic nerve.[1,9,10]

Histologically, most schwannomas are recognized by their distinctive alternation of dense fascicular Antoni A areas, spongy Antoni B tissue with small dark nuclei and variably formed Verocay bodies. A collagenous capsule, hyalinized vessels, xanthoma cells and perivascular hemosiderin complete the diagnostic picture. However, the present case showed the typical histological features of schwannoma. Moreover, this case occurred in an unusual location. Immunohistochemically, schwannomas demonstrate generalized strong staining for S-100 protein. Ultrastructurally, the elongated cells of schwannomas are individually surrounded by duplicated basal lamina. The present case showed typical immunohistochemical findings of schwannoma. Meningiomas may be reactive for the S-100 protein but it is often weak and patchy. Meningiomas also show a membranous pattern of immunoreaction for epithelial membrane antigen. Pituitary adenoma was easily excluded by not only the histology of the tumor but also the lack of immunoreactivity for pituitary hormones. Therefore, schwannomas arising in an unusual location such as this case may require immunohistochemical application for the confirmation of the diagnosis.[11]

In T1-weighted MRI scan usually the schwannoma has an isointense or hypointensesignal compared to brain cortex as like pituitaryadenoma. The schwannoma in T2-weighted image showsmore frequently hyperintense signal than pituitary adenoma. However, it is possible for pituitary adenoma to have variable findings including hyperintense signal which is notdifferent from usual schwannomas. Thus, MRI findings couldnot provide information favouring schwannoma. So, the clinical and radiological presentationof intrasellar schwannomas is consistent with the findingsof pituitary adenomas. Therefore, many surgeons chosetranssphenoidal approach as initial surgical procedure but Wilberger recommended that a trans-sphenoidal approach should be avoided if a schwannoma is suspected preoperatively asschwannomas are fibrous, highly vascular andtumor removal is often incomplete.[4]

Schwanncells are not normally found in the central nervous system and the origin of primaryintrasellar schwannomas remains unclear. Several histopathological hypotheses have been proposed. Russell and Rubinstein suggested an embryonic basis of origin, i.e. the possibility of ectopic Schwann cells resting within the sella.[12] At the time of neural tube closure, neural crest cells divide in the midline, migrate laterally and become segmented into cell clusters between the neural tube and the somites. The neural crest cells migrate widely in the whole body and undergo various differentiations in different tissues. Based on this embryologic process, these Schwann cell nests pinched off in the segmentation of the neural crest are probably left in the intracerebral pituitary gland, leading to the source of sellarschwannomas. Perivascular Schwanncells in the pituitary are another possible source according to some authors. The pituitary is an endocrine organ and has a rich vascular supply. Thus, stimuli evoking proliferation of perivascular Schwann cells in the pituitary may form

schwannomas.[13,14] Also, it was suggested that schwannomas with a tight attachment to the dura may arise from Schwann cellsensheathing the small nerve twigs that innervate the dura. It is supported by the fact that the supplying nerve twigs covering the pituitary are branches of the trigeminal nerve and histologic similarity exists between the mesodermal cells of the pia mater and the neuroectodermal cells having derived from the neural crest. Despite the existence of several theories, none has been proved yet.

CONCLUSION

Schwannoma is an extremely rare tumor inthe sellar and suprasellar region originating from the optic nerve. Sellar schwannoma may show histologic findingsdifferent from conventional schwannoma at other sites. Sellar schwannoma should be included in the preoperative differentialdiagnosis of sellartumor, although the incidence of intrasellar schwannoma is quite lowin this location. Because of thesimilarity with pituitary neoplasms in the clinical and radiologicalaspects, it should be included in the differential diagnosisof sellar and suprasellar lesions.

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Conflicts of interest: None.

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