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Concurrent meningioma and schwannoma; An uncommon presentation of common tumors

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Introduction

Meningiomas and schwannomas are relatively common tumors of the nervous system, however, concurrent tumors are rare. These concurrent tumors are usually associated with Neurofibromatosis type 2, Von Hippel-Lindau disease and sometimes with radiation therapy [1-3].

Several mechanisms underlying the development of these concurrent tumors have been proposed including metaplastic change and bidirectional differentiation. However, the precise pathogenesis is unknown [4]. Only 9 cases of such concurrent tumors have been reported till date. Here we report a case of a 55 year old male patient presenting with a space occupying lesion at C1/C2 level.

Case presentation

A 55 year old male patient presented with back pain and quadriparesis, which developed gradually over a period of 4 months. On examination, no other nodules or spots were identified suggesting any syndromic association. No family history of Neurofibromatosis or Von Hippel-Lindau disease was present. There was no history of prior radiation therapy. Magnetic resonance imaging revealed a dumbbell shaped space occupying lesion with intradural and extradural components as well as mixed signal intensity (Figure 1). Clinical and radiological differential of neurofibroma was raised. The tumor was removed via C1 C2 laminectomy. Peroperatively, the tumor was attached to dura on one side and C2 nerve root on the other.

The specimen was sent for histopathological examination to our laboratory and grossly comprised of multiple fragments measuring approximately 2 cm. Sections taken from the tumor revealed a neoplastic proliferation composed of two definite components. First component showed proliferation of spindled to wavy cells arranged in vague hypocellular and hypercellular areas with verocay body formation. The second component showed whorls of meningothelial cells with abundant intercellular collagen and scattered psammoma bodies (Figure 2A, 2B and 2C).

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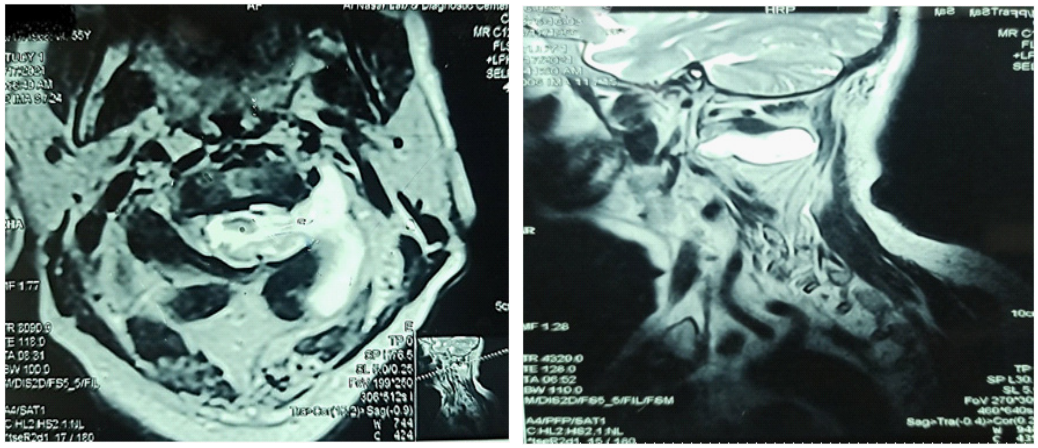

Figure 1: Magnetic resonance imaging studies done preoperatively showing a dumbbell shaped neoplasm with intradural and extradural components.

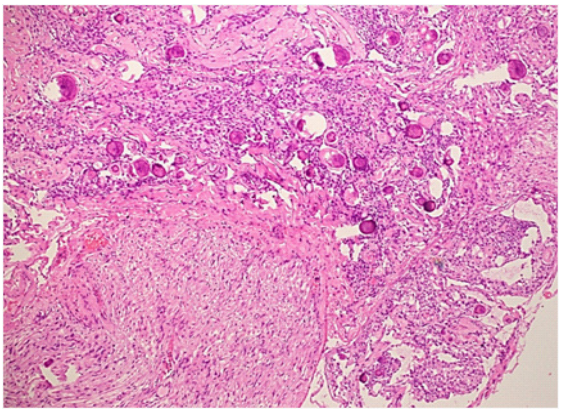

Figure 2A: Low power view displaying two distinct components. Lower portion displays a spindle cell proliferation and upper portion shows proliferation of relatively round cells present in the form of nests and whorls with abundant psammoma bodies.

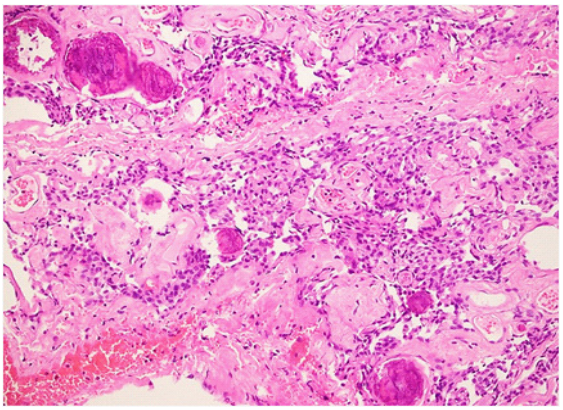

Figure 2C: High power view of the second component composed of meningothelial cells arranged in the form of whorls. Cells have moderate eosinophilic cytoplasm, indistinct cell membranes and round, uniform nuclei. Scattered psammoma bodies are also noted.

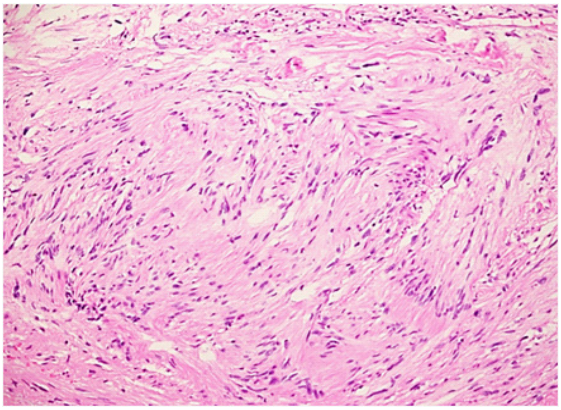

Figure 2B: High power view of the Schwannoma component. Typical Antoni A type areas exhibiting nuclear palisading around the fibrillary processes also known as Verocay body formation characteristic of schwannoma.

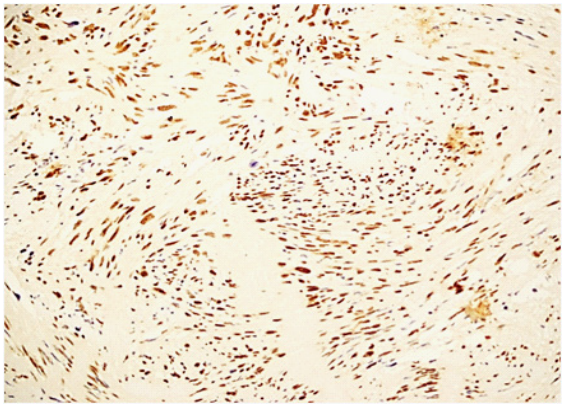

Figure 3A: Diffuse nuclear staining of SOX 10 in the Schwannoma component.

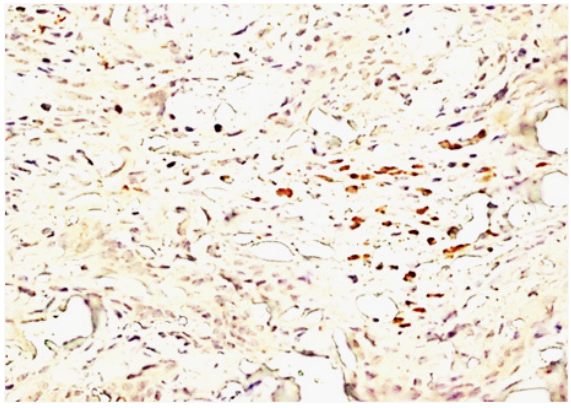

Figure 3B: SOX 10 nuclear staining in the Schwanoma component while meningioma cells are negative.

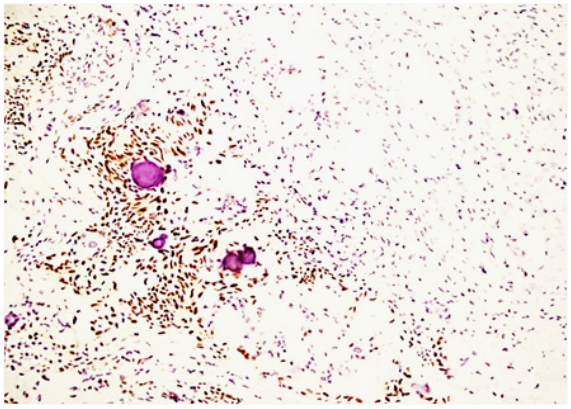

Figure 4A: PR immunostain which stains the nuclei of meningioma while Schwannoma component is negative.

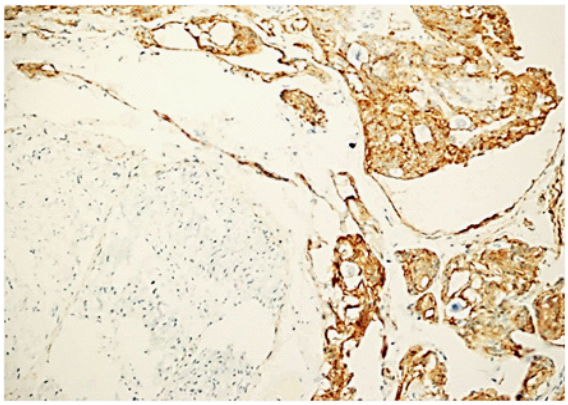

Figure 4B: EMA staining the meningioma component and sparing the Schwanoma component.

No definite necrosis, mitosis or high grade features were noted in the submitted specimen. Due to rarity of this diagnosis, immunohistochemistry was also performed for confirmation of the diagnosis despite unequivocal histological features. Sox 10, which is a very specific and sensitive marker of neural differentiation showed strong nuclear staining in the schwannoma component (Figures 3A and 3B). EMA and PR bothhighlighted the meningioma componentonly (Figures 4A and 4B). The patient had an uneventful post-operative course.

Discussion

The simultaneous development of two tumors at the same location is very rare and a few cases have been reported at the cerebellopontine angle and cervical spine, mostly in association with Neurofibromatosis (NF) or prior radiation therapy. NF2 is a rare disorder affecting about 1 in 40000 newborns and has a autosomal dominant inheritance [4].

Although unclear, multiple underlying mechanisms have been proposed for this rare occurrence [4,5]. One hypothesis is that meningothelial hyperplasia or reactive meningeal changes develop in the vicinity of a Schwannoma [4,6]. Other proposed mechanisms include collision of two separate tumors, metaplastic change in one original tumor or multi directional differentiation of the same progenitor cell into different neoplastic components [4,7,8]. Schwannomas are neuroectodermal tumors that are assumed to originate from Schwann cells of dorsal nerve roots in the subarachnoid space, hence, forming dumbbell shaped tumors along the nerve roots [4,7,8]. Meningiomas are mesodermal neoplasms that are supposed to arise from the arachnoid membranes and that is why almost 90% are present in an intradural location [4]. Some authors believe that schwannoma and meningioma cells arise from the same mesenchymal cell [8-13]. Another hypothesis involves the exposure of two separate neoplastic cell types at the same location as a result of same oncogenic stimulus, with consequent development as a composite tumor [12-15].

Although, MRI is the best preoperative imaging technique for the evaluation of neurological neoplasms, the differentiation between mixed and composite tumors remains a challenging task because both tumors are homogenously contrast enhancing [15]. Preoperative extensive imaging analysis is fundamental as the surgical treatment approachvaries depending on the type of the tumor [14].

Conclusion

In conclusion, our case had no prior radiological or clinical suspicion of a composite tumor. However, histology alongwith immunohistochemistry was quite convincing. Gross total resection was achieved via laminectomy and patient had an uneventful postsurgical course.

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