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Case Report

Cervical intradural extramedullay vascular schwannoma in a thirty eight year old male

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ABSTRACT

Schwannomas are benign peripheral nerve sheet tumours that may arise almost anywhere in the body but are commonly seen in the head, neck region and in the extremities. They may be associated with variable clinical presentations depending on their location. The peripheral nerves are closely related to vascular tissues morphologically and physiologically and therefore schwannomas may be associated with vascular changes like vascular hyperplasia and vascular dilation. Here authors represent one such case where a 38-year-old patient presented with a cervical swelling which on histopathology was diagnosed as vascular schwannoma.

Keywords: Cervical swelling, Schwann cells, Soft tissue tumours, Vascular schwannoma

INTRODUCTION

Schwannomas are benign nerve sheet tumours which originate from differentiated schwann cells. These cells originate from neural crest cells and they surround the peripheral nerve fibres.[1]

Ninety percent of schwannomas are sporadic, five percent of these tumours are associated with meningiomatosis with or without neurofibromatosis type 2, three percent of these tumours are associated with neurofibromatosis type 2 and two percent are associated with schwannomatosis.

Peripheral nerves are closely related to vascular tissues both histologically and physiologically. Schwannoma of peripheral nerve origin is also speculated to be associated with vascular tissues, however, the histogenesis about the vasculature is not fully understood.[2] Therefore, vascular proliferation may be seen in these tumours, but these vascular schwannomas are not very common.

MRI has become an important diagnostic modality in the assessment of spinal tumours. Schwannomas are Iso or Hypo intense on T1 weighted images and hyper intense on T2 weighted images.[3]

Histopathology is the gold standard for the diagnosis of these lesions.

CASE REPORT

A 38 year old male presented to neurosurgery department with a painless neck swelling since 2 years. The mass measured approximately 4×4×3 cms in size. It was firm and non-tender. Overlying skin appeared to be normal. It was painless and was not associated with any other symptoms. There was no significant past history and family history.

MRI spine suggested of an intradural extramedullary space occupying lesion showing contrast enhancement. The patient was taken to OT and transcervical excision of tumour was done under general anesthesia.

Postoperatively the patient did not develop any motor or sensory abnormality. The resected mass was sent for histopathological examination.

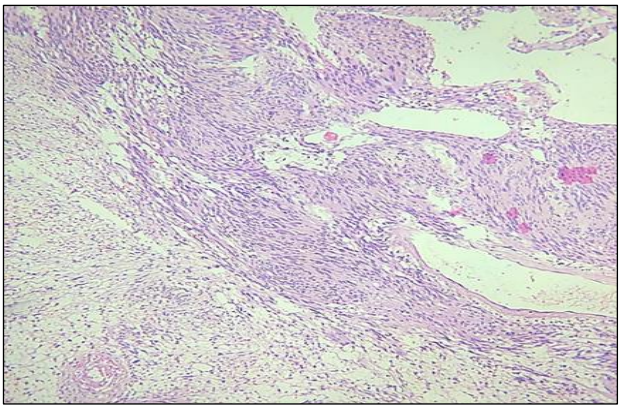

Figure 1: Tumour showing Antoni A and Antoni B areas (H&E 10X).

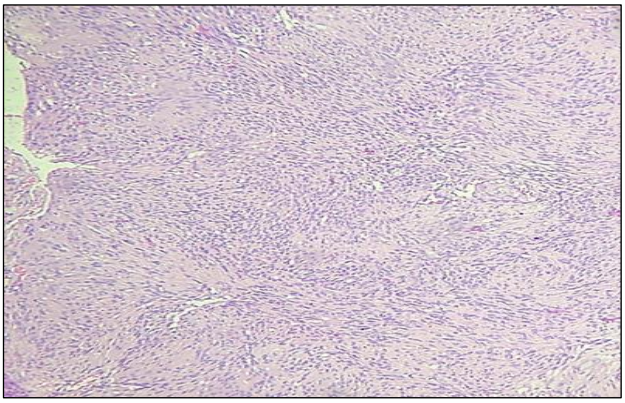

Figure 2: Antoni A areas were composed of spindle shaped cells with elongated nuclei (H&E 20X).

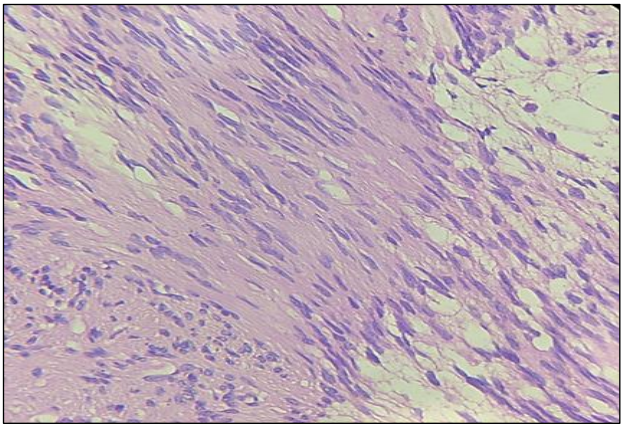

Figure 3: Tumour cells showing nuclear palisading and verocay bodies in antoni A areas (H&E 40X).

Histopathological examination showed presence of an encapsulated tumour showing hypercellular (Antoni A) and hypocellular (Antoni B) areas (Figure 1). The Antoni A areas were composed of spindle shaped cells with elongated nuclei (Figure 2). Mitotic figures were not seen. Nuclear palisading and verocay bodies were prominent (Figure 3). Dilation and hyperplasia of the vessels were observed in both Antoni A and B areas (Figure 4). A final diagnosis of vascular schwannoma was made.

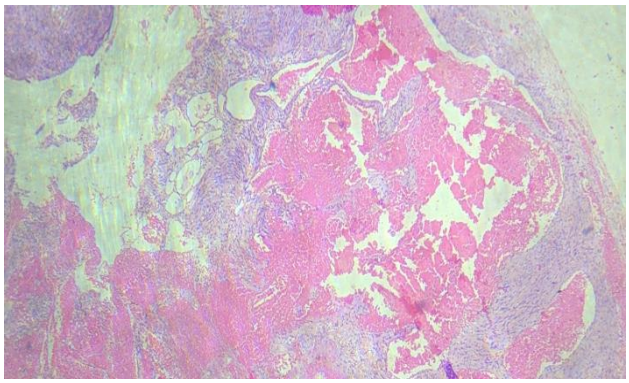

Figure 4: Dilated vascular channels seen in both Antoni A and Antoni B areas (H&E 10X).

DISCUSSION

Schwannomas of head and neck region are not uncommon but they are rarely reported. These tumours are benign, slow growing, encapsulated tumours attached to nerves. They are commonly associated with degenerative changes like cystic degeneration and haemorrhagic necrosis. These changes are not seen in neurofibromas.

The peripheral nerve tissue is closely related to vascular tissue and therefore presence of vascular proliferation may be seen associated with schwannomas of peripheral nerve origin. Growth of these blood vessels in these neoplasms reflect the physiological dependency of nerves on oxygen and nutrients. Eichmann et al suggested that axon guidance influenced vascular network formation and growth.[4]

These vascular schwannomas are very rare. Variously sized blood vessels with dilation and hyperplasia were also noted in our case.

Schwannomas can arise from any site but they are more common on the head, neck and extremities with a predilection to the upper limbs, where they can arise from small or medium sized nerves. Deeply seated schwannomas are mainly seen in the posterior mediastinum and in retroperitoneum.

They can arise from the dorsal nerve roots and these tumours comprise 30% of primary intraspinal tumours. They are characteristically seen in age group of 40-60 years.[5]

The signs and symptoms of these lesions are not very specific and depend on the size of the tumour and its location. The size of the tumour may range from few millimetres to over 24 cms. In our case the size of the tumour was 4×4×3 centimetres.

Majority of patients present with a painless mass but pain may be present in some cases. Other symptoms may include difficulty in breathing (nose), dysphagia (pharynx), epistaxis (nasopharynx), hoarseness (larynx) or it may just present as a painless swelling in there neck (parapharyngeal space).

The preoperative diagnosis of head and neck schwannomas may pose difficulties. Most routine investigations are of limited value in diagnosing these lesions. FNAC helps in distinguishing benign and malignant soft tissue tumours but has low accuracy in characterization of soft tissue tumours especially tumours of neural origin. Histopathology is the gold standard for diagnosis of these cases especially the cases which show uncommon features like vascular dilations and hyperplasia. These cervical spinal schwannomas are benign neoplasms and they usually have excellent prognosis after surgical removal. The risk of recurrence is reported to be less than 10% after complete surgical removal.[6]

CONCLUSION

Schwannomas may be associated with vascular changes like vascular dilatation and hyperplasia due to close relationship between vascular tissues and peripheral nerve tissues. Pathologists should keep in mind that benign schwannomas may be associated with these vascular changes so as to prevent wrongly diagnosing a case of schwannoma as a vascular tumour.

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