

Case Report

Ancient schwannoma in posterior triangle of neck: a rare tumor case report and literature review

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ABSTRACT

Ancient Schwannoma is a rare variant of Schwannomas, a type of benign nerve sheath tumor arising from Schwann cells, hence giving it its name Schwannoma. The term ancient represents the tumor undergoing degenerative changes such as calcification, hyalinization, hemorrhage, cystic degeneration and necrosis and characteristically the loss of Antoni type A tissue. This case report describes a patient having an ancient schwannoma and the management done. Detailed management and findings starting from history and extending to USG, FNAC, CT scan, MRI and excisional biopsy have been described with positive findings supporting the diagnosis like presence of degenerative material in FNAC, peripheral enhancement on CT with the classical Target sign also visible on MRI. The following case reports helps to show how to connect the dots of different findings that may start from the patient's basic history to eventual confirmation by biopsy.

Keywords: Schwannoma, Nerve sheath tumor, Neck swelling, Benign, Neurilemmoma, Peripheral nerve tumor

INTRODUCTION

Schwannomas, also known as neurilemmomas and axonal intraneural Schwann cell tumors, are benign neoplasms of the nerve sheath originating from Schwann cells.¹ Ancient schwannoma is an uncommon schwannoma variant with a course typical of a slow-growing neoplasm.² They were originally described by Ackerman and Taylor in 1951 in a review of 48 neurogenic tumors of the thorax.³

A type of long-standing soft tissue tumor, they are a rare entity and undergo degenerative changes, calcification, hemorrhage and pleomorphism and hence they are commonly misdiagnosed as being a malignant tumor. This case report describes a patient with ancient

schwannoma of the left supra clavicular nerve arising from the neural foramina at C5-C6 level.

CASE REPORT

A 42-year-old woman presented to the general surgery department of GMERS medical college and hospital Gotri in Vadodara with a slow growing visible and palpable mass on the posterior aspect of the left side of her neck for 8-9 months. There was no history of trauma, weight loss or loss of appetite. On examination a single firm mass was noted to be of 5x3x2 cm in size and situated in the posterior triangle of the left side of the neck. On local examination it showed mild tenderness and was mobile in nature. There were no palpable lymph nodes. The external appearance of the swelling from the anterior and lateral view is shown in (Figure 1-2).

USG and FNAC

On Ultrasonography of the local site of swelling, the lesion was found to be hypoechoic along with coarse heterogenous echotexture and minimal internal vascularity. Plain Radiography showed a non-calcified soft tissue mass like opacity with no attachment to the surrounding bones. Next, fine needle aspiration Cytology was performed and 1 Papanicolaou (PAP) stain, 1 Giemsa stain and 3 Hematoxylin and Eosin (H&E) stain were performed on the aspirate. One small cluster of cellular degenerated material was visible under microscope along with blood.



Figure 1: Anterior view of swelling present at lateral aspect of left side of neck.



Figure 2: Lateral view of swelling at the lateral aspect of left side of neck.

CT report

Subsequent CT scan was done and enhanced spiral sequences of neck were obtained from the level of clavicle to the base of skull. It portrayed the presence of a well-defined, spindle shaped, heterogeneously hypodense lesion of size 32x51x42 mm with few necrotic areas. It showed predominant peripheral heterogenous enhancement with few central non enhancing areas within. It caused left neural foramen widening at C6

level. Subsequently MRI neck was also advised for further detail and relations of the swelling.

MRI report

MRI neck was then done using T1, T2 axial, T1 and STIR coronal and T1 & T2 sagittal sequences. Post contrast T1W images were also taken in all three planes. The lesion appeared to be homogeneously hypointense in T1W images, while appearing heterogeneously heterodense on T2W and STIR images.

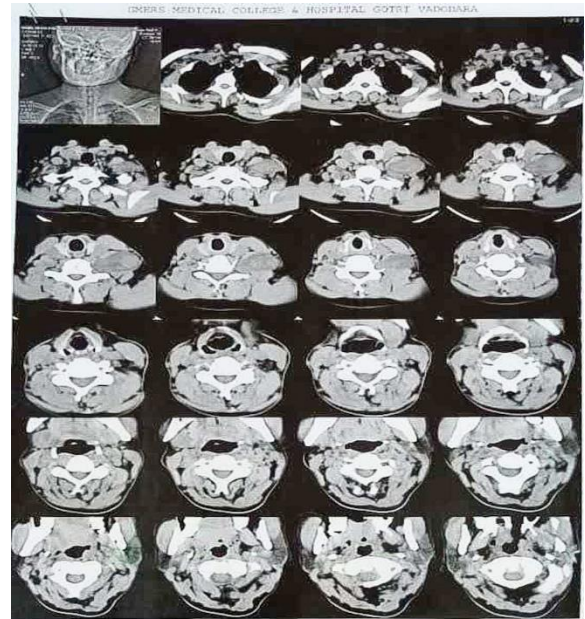


Figure 3: CT Scan slices from level of clavicle to base of skull.

It had a heterogenous thick peripheral enhancement with a central necrotic area. MRI confirmed the origin of the lesion from the left neural foramina at C5-C6 level, along with MODIC type 1 endplate changes at the same. The classical “target sign” was also visible. After appropriate pre-operative investigations and pre-anaesthetic check-ups, an excisional biopsy was scheduled with the clinical suspicion of a peripheral nerve tumor. The routine blood investigations are mentioned in (Table 1). Following all pre operative procedures, investigations and consent the patient was undertaken for Surgery under General Anaesthesia under ASA-2.

Table 1: Routine blood investigations.

Parameters	Observations	Units
Haemoglobin	11	g/dl
RBC	4.17	Mill/cumm
WBC	10600	/cumm
Platelet	366000	/cumm
Creatinine	0.6	Mg/dl
SGPT	15	U/l

Subsequently excisional biopsy was performed.



Figure 4: Lateral slices of CT scan.

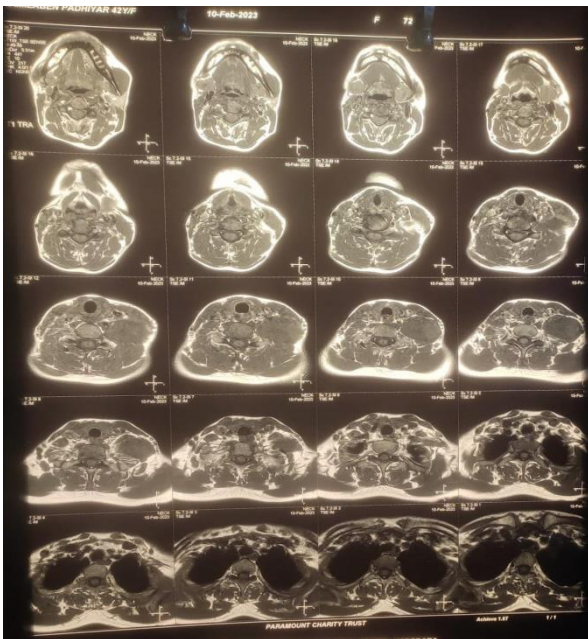


Figure 5: MRI Scan with vertical slices.

The lesion had the following relation to the surrounding tissues: Anteriorly it was compressing the anterior scalene muscle and the posterior aspect of the sternocleidomastoid muscle. Posteriorly it was compressing the middle scalene muscle. The muscles were then retracted and further dissection was done. Inferiorly it was in proximity to the left subclavian artery and laterally was compressing the external jugular vessels. Medially it was abutting the left vertebral artery and posterolaterally was related to the C5 vertebral body. Grossly, the lump was approximately 3x5x4 cm and had

a glossy and smooth surface, which was subsequently cut revealing a central fibrous and necrotic area with a thick capsulated periphery, as shown in (Figure 9).

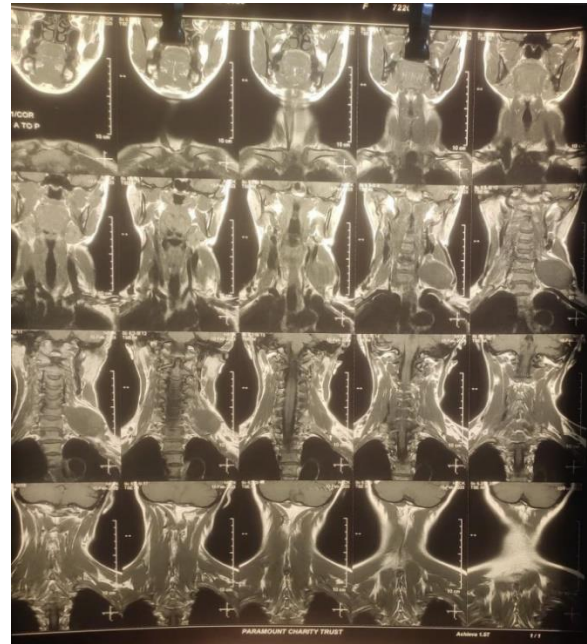


Figure 6: MRI Scan with Coronal section images.



Figure 7: Draping and proper exposure of operative field.



Figure 8: Intra-operative image of lesion.

Excisional biopsy analysis

The tumor was then sent for analysis by preserving it in formalin. The histopathological findings reported a well circumscribed encapsulated mass consisting of hypercellular and hypocellular areas and focal area of Verocay bodies. The cells were narrow, elongated and wavy with tapered ends, interspersed with collagen fibres, suggestive of Antoni Type A cells. The hypocellular areas had pink eosinophilic acellular material. Areas of cystically dilated hyalinized blood vessels and degenerative type of cytological atypia also was seen. All the above findings confirmed the suspicion of the tumor being an ancient schwannoma.

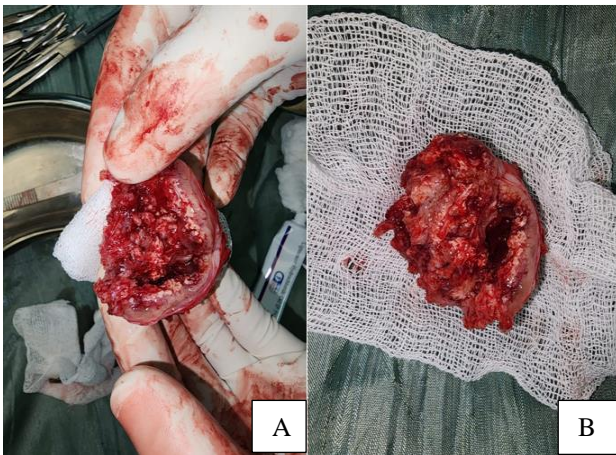


Figure 9 (A & B): Immediate post-operative cut section of the lump.

DISCUSSION

A schwannoma is a common benign nerve sheath tumor. These tumors can arise from any nerve covered with a Schwann cell sheath, which include the cranial nerve (except for optic, olfactory), spinal and autonomic nervous system.⁴ Ancient schwannoma commonly occurs in the head and neck, thorax, retroperitoneum, pelvis, and extremities. They're mainly seen in the middle and elderly age, with no sexual predominance, and are usually asymptomatic to start with, but the patient may develop symptoms based on the location of the tumor, predominantly due to exertion of pressure. The development of a schwannoma in the posterior triangle of neck can be easily mistaken for lymphadenitis, lipoma, dermoid cyst, secondaries in the lymph nodes and so on. The diagnosis can be made by investigations like ultrasonography, FNAC, CT scan/MRI or a Biopsy. Schwannoma is a slow-growing benign tumor and can be a large tumor with degeneration, especially when the tumor is situated in the deep regions such as the mediastinum and retroperitoneum.⁵ Schwannoma with pronounced degenerative changes is known as an ancient schwannoma, which is a rare variant of schwannoma and is usually a deeply situated large mass of long duration.^{5,6} Since these changes were noticed in patients with a long-

standing duration of the tumor, it was given the term "ancient schwannoma". The classical characters of it being cystic necrosis paired with increase in cellularity. Additionally diffuse areas with fibrosis, increased deposition of matrix, hyalinization, ectatic vessels with thrombus within them, hyperchromasia and nuclear pleomorphism may be visible microscopically. In the case of our patient the clinical picture and FNAC findings supported the diagnosis of an ancient schwannoma. Complete surgical excision of the tumor is the treatment of choice, because these are radio-resistant tumors. During excision of the tumor, all efforts should be made to preserve the nerve but it is difficult because of its dense attachment to the tumor. Incomplete excision may result in slow recurrence over months to years. Recurrence and malignant change of ancient schwannoma are very rare but some cases have been reported. The diagnosis is confirmed after histopathological examination. Tumor cells are usually positive for S-100 antigen in immunohistochemical examination.

CONCLUSION

In conclusion a case of ancient schwannoma in the left supraclavicular region is described, showing it can be insidious in onset and easily mistaken for another neck swelling of lymph nodes, lipoma, etc. The initial or presenting symptoms depend on the site of the lesion and the nerve that is involved and hence special attention should be given to them. It should also not be left out in the differential diagnosis of swellings especially when degenerative signs like cystic degeneration, calcification or necrosis are present on initial investigations like FNAC or USG. Differentiating clinical features, investigations and radiological signs like target sign should be noted. The histopathological findings after biopsy like the presence of Verocay bodies, degenerative cellular material with a positive Immunohistochemical S-100 stain aid and helped confirm the diagnosis post excision.

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