BRACHIAL PLEXUS SCHWANNOMA

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ABSTRACT

Brachial plexus schwannoma is a rare entity representing only 5% of schwannoma. They create a great challenge to surgeons due to their rare occurrence and complex anatomical location. It is a benign nerve sheath tumor and the commonest presentation is painless slow growing supraclavicular firm swelling. Infiltration or pressure over the surrounding structure might give pain, paraethsia etc. One of the differential diagnosis of such swelling should be schwannoma. MRI is helpful in diagnosis since FNAC may damage the nerve and hemorrhage can give rise to pressure symptoms. Complete excision of the lesion along with preservation of the nerve is needed.

Keywords: Brachial plexus schwannoma, excision, FNAC, MRI

INTRODUCTION

Brachial plexus schwannoma is a rare entity representing only 5% of schwannoma. They create a great challenge to surgeons due to their rare occurrence and complex anatomical location. Commonest presentation is a painless slow growing supraclavicular swelling. Some may present with pain, tingling and paraesthesia due to infiltration or mass effect. MRI is helpful in diagnosis of this condition. One of the differential diagnosis of such swelling should be brachial plexus schwannoma. We present a rare case of brachial plexus schwannoma presenting as a slow growing supraclavicular swelling.

CASE REPORT

Thirty-one-year-old female presented with history of heaviness left upper limb for three years, which increased on strenuous work. She presented with painless swelling in left supraclavicular region for 6 months which had gradually increased up to present size. There was no weakness, tingling, paresthesia and wasting of limb. There was no significant past medical and family history. On examination there was a 5cm x 5cm firm, globular swelling in the left supraclavicular region with normal overlying skin, and not mobile in vertical as well as horizontal direction. There was no tingling sensation on palpation of the swelling. There were no other swellings, hamartoma, neuroma, café au lait spots. On nasopharyngolaryngoscopic (NPL) evaluation bilateral vocal cords were normal and mobile. Ultrasonography, CT scan and FNAC from the swelling was done. Radiologically there was well defined hypodense lesion however, FNAC was inconclusive. MRI showed well defined significantly enhancing lesion in prevertebral space extending from C7, D1 and D2 (Fig 1).

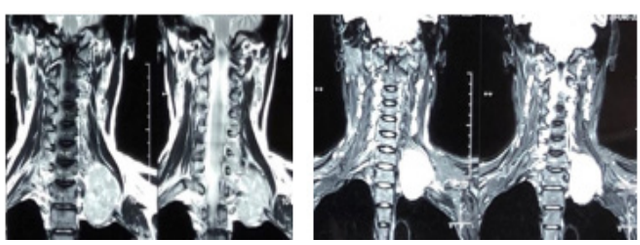

Fig 1: T1 and T2 image of MRI showing lesion in left paravertebral region at level of C7, D1 and D2 vertebral bodies

Lesion was completely excised with supraclavicular horizontal incision under general anesthesia. Per operatively tumor was adhered to the C6,7 nerve root which was stretched and thinned. The tumor was dissected off the tumor completely (Fig 2).

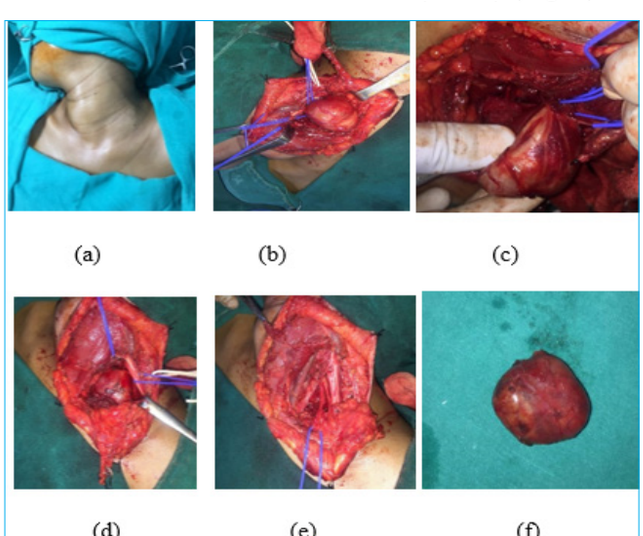

Fig 2 (a-e): Per operative finding during excision of tumor, 2f - Gross specimen after surgical excision

Grossly the specimen was oval in shape, measuring 4.5cm x 3.5cm x 2.5 cm encapsulated soft tumor. The cut surface was yellowish with areas of hemorrhage. The histopathological diagnosis was schwannoma. Histologically, it consisted of Antoni A and Antoni B cells with no atypical cells and no necrosis (Fig 3). Focal pseudocystic areas and foamy histiocytes were also found, as well as hyalinized blood vessels. There was no nuclear atypia or mitotic activity. Immunohistochemical analysis showed diffuse strong expression of S-100 protein and glial fibrillary acidic protein (GFAP) in tumor cells. Postoperatively, the patient developed paresthesia and numbness in the left thumb and forefinger which completely improved on eight post-operative day. Power in the operated limb was 5/5 with intact sensation.

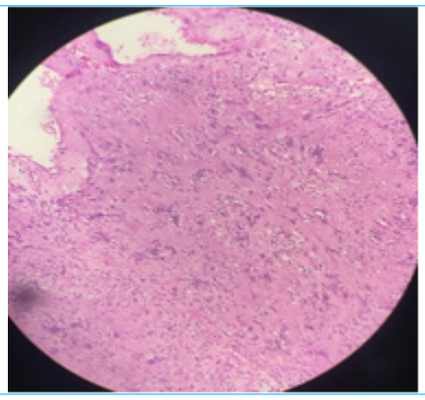

Fig 3: Final histology showing spindle shaped cells arranged in interlacing fascicles with nuclear palisading

DISCUSSION

Schwannoma or neurilemmoma is a benign encapsulated nerve sheath tumor that arises from the schwann cells along the course of a nerve and can affect the third to twelfth cranial nerves, peripheral and autonomic nerves.[1] Brachial plexus schwannoma is a rare entity and represent only 5% of schwannoma. They create a great challenge to surgeons due to their rare occurrence and complex anatomical location.[2] Other more common nerves to arise schwannoma in the neck are vagus and cervical sympathetic chain. Head and neck schwannoma represent 25% of schwannomas and vestibular schwannoma is the commonest. Generally schwannoma arise sporadically as a benign, single swelling.[3] Small tumors are usually uninodular but larger tumors may be multinodular with degenerative features including cyst formation, fibrosis and calcification.[4,5] Clinical presentation of brachial plexus tumors is variable according to its location, extension, neural elements involved and pathology. Symptoms are caused by direct nerve invasion, infiltration of surrounding tissues, or local mass effect painful paresthesia in the distribution of the involved nerve induced by percussion is the single most useful sign in diagnosing schwannoma.[4] Positive Tinel's sign carries a high predictive value for schwannoma.[6]

It may be associated with Von Recklinghausen disease which has multiple neuroma, hamartoma, café au lait spots. Differential diagnosis of slowly growing painless supraclavicular mass can be metastatic lymph node, hamartoma, tubercular lymphadenopathy, sarcoidosis etc. Consideration must be of brachial plexus schwannoma. Tingling sensation in the upper limb on percussion of the swelling is diagnostic of brachial plexus schwannoma. Symptoms are due to infiltration of nerve fascicles or pressure effect. Long standing cases might have loss of sensation and muscle atrophy. Diagnosis is done based on clinical and imaging studies. FNAC should be avoided to prevent injury to the nerve fascicles or avoid pressure effect due to hemorrhage.

Treatment is complete excision or enucleation of the tumor with preservation of the nerve. Histologically, the Antoni type A and type B is used to describe the growth pattern of tumor.[2] Resection of tumor is the treatment of choice in most of benign and malignant brachial plexus tumors. Treatment options includes expectant observation in asymptomatic patients, surgery for better long-term results in patients with progressive or symptomatic disease, and radiotherapy in symptomatic patients unsuitable to undergo surgical treatment.

According to Knight et al., mass arising from a nerve trunk which causes pain and is attended by deepening loss of function is considered a malignant tumor until proven otherwise.[4] An anterior supraclavicular approach is convenient for tumors involving roots and trunks. Post-operative neurological dysfunction may occur after total resection. Tang et al. have reported in their study that schwannomas had in large proportion fascicular involvement that would consequently lead to distal neurological deficit because fascicular involvement could not be identified preoperatively.[6]

CONCLUSION

Brachial plexus schwannoma is rare nerve sheath tumor which should be considered in patient presenting with slow growing asymptomatic supraclavicular swelling. MRI is helpful in diagnosing the case. FNAC can be avoided for the diagnosis to prevent injury to the nerve and surrounding structures. Complete surgical excision of the lesion with preservation of nerve is mainstay treatment.

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