Research Article

Schwannoma Arising at Unusual Locations: A Report of 4 Cases

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Abstract
Neurilemmoma (schwannoma) is a benign, encapsulated perineural tumor of neuroectodermal derivation that originates from the Schwann cells of the neural sheath of motor and sensitive peripheral nerves. Schwannomas are commonly seen in the head and neck region, flexor surface of extremities. However, schwannoma can arise from any cranial or spinal nerve as a globoid encapsulated tumor. We report a case series of four cases of schwannoma arising from four unusual sites one presenting as a parotid tumor; second one as a pleural mass, third as parapharyngeal mass and the fourth one as a nasal mass.

Keywords: Schwannoma; parotid, pleural, parapharyngeal, nasal mass

1. Introduction
Neurilemmoma (schwannoma) is a benign, encapsulated perineural tumor of neuroectodermal derivation that originates from the Schwann cells of the neural sheath of motor and sensitive peripheral nerves. The etiology is still unknown. In 1910, Verokay first described a group of neurogenic tumours; he referred them to as 'neurinomas'. In 1935, it was proposed that these tumours arose from nerve sheath elements and they were termed 'neurilemmomas.' Of the large variety of terms under which these tumours have been reported in the past, only three are still in current use: neurinoma, neurilemoma (neurilemmoma), and schwannoma. Both electron microscopy and immunohistochemistry have established that these tumours are composed of Schwann cells.

Schwannomas may arise from cranial and spinal nerve roots or from peripheral nerves. Schwannomas have a predilection for sensory nerves. The vast majority of intracranial schwannomas arise from the eighth cranial nerve. Of the motor nerve, facial is most frequently involved. Spinal tumours arise predominantly from posterior roots.

2. Case Series
2.1 Case Report I: (parotid schwannoma)
A 25 year old man presented with swelling and pain at the parotid region for the last 1 year. CT Scan revealed right sided parotid mass (fig.1A, B).

Fig. A: CT Scan- rt. parotid mass

Subsequently, Parotidectomy was done and on gross examination there was a well circumscribed globular encapsulated mass measuring 2.5 cm in diameter along with salivary gland tissues (fig.1). On cut section, it was homogenous, solid & greyish white (fig.2)
Microscopic examination revealed a tumor mass composed of spindle shaped cells with nuclear palisading. Antony A and Antony B - both areas are noted. Verocay bodies are also seen. Diagnosis of parotid schwannoma was made (fig. 3, 4 & 5).

2.2 Case Report - 2 (Schwannoma, presenting as pleural mass)

A 68 year old male patient presented with chest pain, cough & dyspnoea along with pleural effusion on the right hemithorax. A well-circumscribed, round mass that is of homogenous soft-tissue density on chest x-ray and plain CT image was noted (Fig. 6A).

The mass was excised along with the pleural tissue. On gross examination, a well circumscribed, firm, round mass measuring 56mm in diameter was noted. (Figure-6)
Microscopic examination revealed well encapsulated tumor mass composed of spindle shaped cells with nuclear palisading with attached pleura at periphery. Antony A and Antony B both areas are noted, Verocay bodies are also seen. Diagnosis of pleural schwannoma was made (Fig.7, 8& 9).

![Fig.7 well encapsulated tumour with attached pleura](image1)

![Fig.8 L.P. View showing nuclear palisading & verocay bodies](image2)

![Fig.9 H.P. View confirming the same findings](image3)

2.3 Case report- 3(schwannoma presenting as parapharyngeal mass)

A 40 year male patient reported with a swelling in the throat, difficulty in swallowing since last 1 year. She also complained of hoarseness of voice for a period of 2 years.

MRI shows a right parapharyngeal mass compressing the esophagus & trachea (fig.10A).

![Fig. 10A MRI-RT. Parapharyngeal SOL](image4)

Excision of the parapharyngeal mass was done and the gross specimen showed –A globular tissue (4x1.5x1 cm), greyish white in colour. Cut section showed blackish haemorrhagic spot and cystic areas in the centre (fig.10, 11).

![Fig.10 oval solid parapharyngeal swelling](image5)

![Fig.11 cut section showing central cystic areas](image6)

H/P examination showed the well encapsulated spindle cell tumour with nuclear palisading with Antoni A & Antoni B areas & Verocay bodies. Thick hyalinised blood vessels with inflammatory cell and foam cells with areas of haemorrhage were also seen (fig.12, 13, 14). Complete excision was achieved without any complications. Postoperative recovery was un-eventful and no recurrence was seen.
2.4 Case Report -4(Schwannoma, presenting as recurrent nasal mass)

A 18 years old female presented with progressive nasal obstruction, polypoidal mass in right nasal cavity, swelling at the right side of the nose near inner canthus of right eye, and headache for last 8 years. Past history of swelling at the right side of nose at the age of 9 years. Operation was done then in our hospital and diagnosis of nasal neurilemmoma was made. After 1 year of operation, she again developed mass in the right nasal cavity which was increasing in size gradually. And at the age of 18 years, she again came to this hospital. In between that period, no intervention was done. On examination there was right sided proptosis and a large mass filling the right nasal cavity.

X-ray PNS and recent CT scan showed a large mass in right fronto-ethmoidal region with mass effect on adjacent structure (causing right sided proptosis) and mild right maxillary sinnitis (fig.15A,15B). Chest X-Ray and other haematological examination were within normal limit. Excision of the mass was done and sent for histopathological examination.

On gross examination, multiple greyish brown tissue pieces with irregular nodular papillary surface. (Figure 15, 16).

On microscopic examination, tumour mass showed respiratory epithelium overlying the unencapsulated tumour composed of spindle shaped cells with nuclear palisading showing both antony A and antony B areas. Verucay body was also noted. The histological feature is consistent with nasal schwannoma. (Figure 17)
3. Discussion

Schwannoma is a very rare mesenchymal tumor of parotid gland. Guzzo et al, reported 8 cases of parotid schwannoma over a period of 30 years. Gross et al reported 15 cases of parotid schwannoma from Mayo Clinic from 1975 to 2010. In another study by Kang et al conducted over 10 years, there were 4 cases of parotid schwannoma among 21 cases of extracranial non-vestibular head neck schwannomas. Shimizu K reported 5 cases of intraparotid schwannomas. Gobindan et al, Shrestha et al, Ali et al reported cases of parotid schwannoma from India.

Parotid schwannoma usually arises from facial nerve and its branches or other nonfacial peripheral nerves running through the parotid gland but facial nerve or its branches are the most common nerve of origin. Preoperative diagnosis of parotid schwannoma is by imaging study is difficult. FNAC is often inconclusive or wrongly diagnosed as pleomorphic adenoma. According to Guzzo et al more than 75% cases of intraparotid facial nerve schwannoma remain unknown until surgery. In our case, the patient was a 25 year male presented with pain & swelling over right parotid region for the last 1 year. Parotidectomy was done after routine investigations Histopathological examination confirmed the diagnosis.

Schwannoma arising from the parotid gland is rare. The lesions are composed of compact spindle cells arranged in short bundles or interlacing fascicles, showing nuclear palisading, and Verocay bodies, formed by two compact rows of well-aligned nuclei separated by fibrillary cell processes. Antoni B areas are composed of compact spindle cells arranged in short bundles or interlacing fascicles, showing nuclear palisading, and Verocay bodies, formed by two compact rows of well-aligned nuclei separated by fibrillary cell processes. In the differential diagnosis of solid, solitary, and well-defined tumors, schwannoma should be included in the differential diagnosis of solitary, solitary, and well-defined pleural tumors.

In our case, we found no features of malignancy. Our case of benign schwannoma of the right hemithorax presented with acute respiratory failure which was unusual. Parapharyngeal Space (PPS) tumours are very rare and account for only 0.5% of all head and neck tumours. Approximately 20% are neurogenic and account for the most common neural tumour next to salivary gland tumour found in the PPS. For 50% of parapharyngeal schwannomas, Vagus nerve is the commonest site of origin. Up to 45% of schwannomas are seen in the head and neck region. The clinical presentation of schwannoma depends on the anatomical area involved. Surgical excision has been the treatment of choice and recurrence is very rare. The PPS is a potential site reported to house plenty of tumours, most of them being benign. Generally schwannomas are characterized by slow and asymptomatic growth; however, its progressive growth in parapharyngeal region may result in pressure effect manifestations like dysphagia and hoarseness of voice. Although schwannoma is most commonly seen in young and middle aged adults, the parapharyngeal schwannoma may be observed in patients between 30 and 70 years of age.

In the present case patient was 40 years old male and suffered from hoarseness of voice for 2 years and difficulty in swallowing for 1 year. Microscopically they are consistently encapsulated with the presence of Antoni A and Antoni B cells along with Verocay bodies. Longstanding schwannomas are known to show cystic degeneration, fibrosis and hemorrhage which were also seen in the present case. Nasal schwannoma is a slowly growing mass. Symptoms depend on the location of the tumor and depend on the location of the tumor and also size.

Histopathologically, most schwannomas are tumour masses surrounded by a fibrous capsule consisting of epineurium and residual nerve fibers. The hallmark of a schwannoma is the biphasic pattern of alternating Antoni A and B areas. Antoni A areas are composed of compact spindle cells arranged in short bundles or interlacing fascicles showing nuclear palisading, and Verocay bodies, formed by two compact rows of well-aligned nuclei separated by fibrillary cell processes. Antoni B areas are hypocellular area shows spindle or oval cells arranged haphazardly in the loosely textured matrix. On occasion, schwannomas develop cystic spaces lined by Schwann cells that assume a round or epithelioid appearance. S-100 protein is strongly expressed by most cells in a schwannoma, in contrast to the cells of neurofibroma, which variably express the antigen. Leu-7 and occasionally glial fibrillary acidic protein is strongly expressed by most cells in a schwannoma.
protein (GFAP) are present in these tumors.25 Unlike other areas nasal schwannoma is usually not encapsulated. Mey et al29 in a case series of five nasal schwannoma and another series of nasal schwannoma cases by buob et al27 showed that none of the cases were encapsulated. According to these authors this peculiarity could be explained by the development of these tumors from sinonasal mucosal autonomic nervous system fibres, which are devoid of perineural cells similar to the case of gastric schwannoma.23,28,33 Our case was also not encapsulated microscopically and probably supporting the theory.

The treatment of choice is complete removal of the tumor and recurrence is extremely rare.37 Malignant schwannoma usually recur but itself is very uncommon.36,37 Our case showed recurrence but the histological feature of malignancy were not there and so the recurrence may be due to incomplete removal. Although rare, nasal schwannoma should be considered as differential diagnosis when solitary nasal mass is encountered.

References
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