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Primary melanotic schwannoma with typical histology

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Primary Melanotic Schwannoma with Typical Histology

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INTRODUCTION
Melanotic schwannomas comprise of unique schwann cells which also secrete melanin pigment and are thought to arise from neural crest cells. Psammomatous schwannomas of the Carney complex and non-psammomatous sporadic tumors are the two specific types. Carney complex is characterized by abnormal skin pigmentation, myxomas of the heart and skin, endocrine cancers and schwannomas. Schwannomas associated with the Carney complex present in the younger age group, compared to the sporadic forms. The most common symptoms of a melanotic schwannoma are pain in the respective dermatome, loss of sympathetic nerve function and cutaneous pigmentation.

CASE REPORT
A 17-year male, non-smoker, presented to the outpatient clinic with pain and discomfort in the right upper chest for 6 months. He had temporary relief from analgesic medications but the pain recurred and gradually increased during the previous 2 months. Patient also complained of diaphoresis on the right upper quadrant of the chest. There were no signs of weakness in the limbs and no complaint of weight loss.

On examination, patient was vitally stable with normal vesicular breathing and audible heart sounds. Central nervous system examination showed no deficits. Systemic examination revealed no visceromegaly or incontinence. Chest expansion was equal on both sides.

Patient had no hindrance in daily activities and his diet was adequate. His primary laboratory investigations were within normal limits.

X-ray revealed a soft tissue mass originating from the midline and abutting the superior lobe of the right lung (Figure 1). A subsequent CT scan displayed well defined, round, solid mass in the right posterior mediastinum with epicenter at the right neural foramina of the T3 vertebra, which was suggestive of a neurogenic tumor (Figure 1). Surgery was planned and patient underwent right sided thoracotomy. Intra-operatively, the mass was found to be adherent to superior segment of right upper lobe, anterior chest wall and the posterior chest wall deep to the spinal canal. Grossly, tissue was solid in consistency and covered in blackish discharge (Figure 2). A wide local excision was performed which was complicated by
a minor injury to the dura matter. Tissue samples comprising of the gross tumor and mediastinal lymph nodes were analyzed microscopically. Patient recovered after the surgery and underwent respiratory physiotherapy. The patient has remained asymptomatic one year postoperatively with no recurrence.

Microscopically, a poorly circumscribed, heavily pigmented tumor was seen which was arranged in multinodular configuration, i.e. composed of large nodules separated by fibrosis. The individual cell morphology was obscured by the abundance of pigmentation and, therefore, required its bleaching. Within these nodules, the tumor cells predominantly revealed epithelioid morphology along with abundant cytoplasm and indistinct cell outlines resulting in a syncytial pattern. Periphery of the nodules showed haphazard fascicles of spindle shaped cells. This dark brown, fine to coarsely granular pigment was mainly intra-cytoplasmic but extracellular spillage was seen. The melanotic nature of the pigment was confirmed by positive staining with Masson-Fontana special stain. After extensive sampling, few foci of psammomatous calcification were also appreciated. The nuclei were oval and vesicular with prominent nucleoli and occasional pseudo-inclusions. Few scattered mitotic figures were also identified. Nerve tissue was also identified entrapped in between as well as at the periphery of the tumor. Tumor was predominantly abutting the adjacent lung tissue along with focal invasion into it (Figure 3).

Interpretation of the immunohistochemical stains was difficult due to similarity of the dark brown chromogen used for immunohistochemistry with melanin pigment. In few areas with limited pigmentation, the tumor cells were sparsely positive for immunohistochemical stains, S-100 and HMB-45.

A possibility of malignant melanoma was also considered; however, contemplation of the age, site of lesion, syncytial pattern of cells, lack of marked pleomorphism or brisk mitotic activity and presence of rare psammoma bodies directed the diagnosis in favour of melanotic schwannoma.5

DISCUSSION

Melanotic schwannomas of spinal nerve root origin are a rare entity but have a 15% mortality rate due to difficult surgical approach and frequent recurrence (18.2%).1,6 Less than 100 cases of melanotic schwannomas have been described, mostly arising from the spinal nerve roots.5-7 Melanotic schwannomas comprise less than 1% of all peripheral nerve sheath tumors.6 Morphologically, it is similar to a malignant melanoma; therefore, it is important to differentiate it microscopically. Melanotic schwannoma has distinct histological features as were seen in our case. Definitive diagnosis is essential as further management is significantly specific for these cases. Melanotic schwannomas are managed through wide local excision and do not require radiotherapy if complete excision is achieved. When complete excision is not possible, postoperative radiotherapy is indicated. Thoracotomy is an ideal approach for such tumors as adequate visualization of the posterior chest wall is required.

The tumor has ambiguous histology owing to features of two distinct cell lines and diffuse hyperpigmentation.3 Malignant melanoma should always be kept as a differential until it is excluded on microscopy and tumor markers.

Latest research reveals that melanotic neural tissue tumors comprise of several mutational and chromosomal aberrations which can reliably be differentiated by methylene profiling.9 Furthermore, literature describes the tumor being resected through minimally invasive techniques using video-assisted thoracoscopy (VATS) but due to the vast local extension plus adherence of the
tumor in this case, a thoracotomy was done to access and resect the complete mass.\textsuperscript{10}

REFERENCES