CASE REPORT

Cellular schwannoma masquerading as malignant peripheral nerve sheath tumour: a diagnostic dilemma

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SUMMARY

We present a case of a 15-year-old girl with a pulsatile, rapidly enlarging mass at the root of the nose suspected to be malignant. Excisional biopsy showed worrisome histological features; however, a final diagnosis of cellular schwannoma was reached excluding the possibility of malignant peripheral nerve sheath tumour by histological and immunohistochemical attributes. Cellular schwannoma, a pseudosarcomatous entity, is a rare benign neoplasm that may cause bone erosion and may be mistaken for a malignancy, clinically and histologically. Diagnosis of cellular schwannoma is essential to prevent mismanagement as it never metastasises and responds to local excision as opposed to aggressive treatment required by a malignant neoplasm.

BACKGROUND

Schwannoma (neurilemoma) is a benign tumour found in the extremities and the head and neck. Cellular schwannoma, on account of its cellularity, is often mistaken for malignant nerve sheath tumour and low-grade malignant smooth muscle tumours.¹

We report a case of cellular schwannoma with typical clinico-histological features of malignant peripheral nerve sheath tumour (MPNST).

CASE PRESENTATION

A 15-year-old girl presented to the surgery outpatients department with swelling at the root of the nose. The patient had pulsations in the mass on straining and coughing and an associated history of weight loss. The swelling was 5×5 cm globular cystic mass arising from deep structures, suspected to be malignant. Excised specimen was sent for histopathological examination which showed a partially capsulated mass with heterogeneous cellularity (figures 1 and 2). The hypercellular areas were worrisome and showed bundles of spindle-shaped cells with bullet shaped and fusiform nuclei (figure 3) having atypia and increased mitosis (20/10 high power field). This led to suspicion of malignant peripheral nerve sheath tumour (figure 4). The cells also showed areas of herring bone-like arrangement giving an impression of fibrosarcoma. However, hyalinised thickened blood vessels and lymphocytes were seen admixed in these areas. In view of the atypical picture the diagnosis was narrowed down to cellular schwannoma and MPNST.

Figure 1 Hypocellular and hypercellular areas along with myxoid areas and blood vessels, H&E, ×100.

However, the mitotically active atypical tumour was devoid of areas of necrosis. The mitoses were also typical. The tumour shared its histological picture with classical schwannoma with hypercellular and hypocellular areas, hyalinised blood vessels and lymphocytic infiltrate; however, it lacked the presence of Verocay bodies. The tumour cells were negative on Van Geison staining (figure 5)

S-100 immunostaining showed strong diffuse positivity (figure 6). A final diagnosis of cellular schwannoma was made after clinical, histopathological and immunohistochemical correlation.

INVESTIGATIONS

The patient underwent an x-ray examination showed no bony erosion; however, there was expansion of ethmoidal and frontal sinuses.

Figure 2 Thick-walled blood vessels and increased cellularity, ×100.

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Figure 3 Increased cellularity, with fusiform and bullet shaped nuclei, $\times 400$.

Ultrasonographic examination showed an echogenic lesion arising from soft tissue around areas of sinuses without the involvement of bone. MRI examination could not be carried out because of financial constraints.

The patient, however, wanted excision; therefore, the lesion was excised and submitted for histopathological diagnosis. Fine needle aspiration cytology and biopsy were therefore not done.

DIFFERENTIAL DIAGNOSIS

The primary differential diagnosis which needed to be excluded was MPNST. Histomorphological and immunohistochemical means helped us to reach a final diagnosis in our case.

DISCUSSION

Schwannoma despite being a benign peripheral nerve tumour may recur if not completely excised. It presents as a rapidly enlarging painless mass arising from small- and medium-sized nerves.¹

Cellular schwannoma is a well-recognised and unusual variant of benign schwannoma and due to its atypical features, may lead to an erroneous diagnosis of malignancy in about 28% of the surgical biopsy and resection specimens.²⁻⁴ First described by Woodruff *et al*⁵ in 1981, cellular schwannoma accounts for approximately 5% of the benign peripheral nerve sheath tumours. It predominately affects middle-aged adults and occurs

Figure 4 Hypercellular areas with increased mitosis, atypia and nuclear pleomorphism, $\times 400$.

Figure 5 Van Geison stain showing negativity, $\times 100$.

mostly as a slow-growing tumour in the paravertebral region of the mediastinum and retroperitoneum.⁶ It may show bone erosion and neurological symptoms caused by the compression of the nerve roots, and is histopathologically characterised by the presence of compact spindle cells arranged into fascicles, variable nuclear hyperchromasia and pleomorphism, lack of Verocay bodies and typical predominance of Antoni A areas, and occasionally increased mitoses and foci of necrosis.^{7,8}

Cellular schwannomas may be quite large in size, with sheets of eosinophilic cells, having bullet-shaped nuclei along with characteristic features of schwannomas, that is, hyalinised thick blood vessels and foamy histiocytes. However, nuclear pleomorphism, atypia and mitosis may be mistaken as MPNST, as seen in our case.

Distinction from other tumours exhibiting spindle cells fascicles (fibrosarcoma, synovial sarcoma, malignant fibrous histiocytoma and MPNST) is somewhat difficult and it has been suggested that the cellular schwannoma should be included in the group of the so-called pseudosarcomas.⁹

The important differentiating points from low-grade malignant smooth muscle tumours or MPNSTs are gross encapsulation, absence of necrosis and alternating hypocellular and hypercellular areas, along with thick-walled blood vessels and histiocytes.¹

Unlike these malignant tumours, cellular schwannoma reveals a strong and diffuse reactivity for S-100 and an unevenly

Figure 6 S-100 stain showing positivity in cellular areas and delineating thick walled blood vessels, $\times 100$.

distributed reactivity for glial fibrillary acidic protein on immunohistochemical study.^{10 11} Furthermore, the clinical course is benign and the recommended treatment is surgical excision.^{12 13}

In conclusion, cellular schwannoma, a pseudosarcomatous entity, is a rare benign neoplasm that may cause bone erosion and reveals histological features of a pleomorphic and hypercellular tumour but gross total resection appears to be the curative treatment yielding favourable results, and no adjuvant therapy being necessary.³ Distinction from malignant tumours is possible if we use strict criteria on histological and immunohistochemical analyses, avoiding occasional mistaken diagnoses.

The outcome is favourable without metastatic spread¹⁴ and this is the principal reason for delineating it as a separate entity. Thus correct identification of cellular schwannoma is essential to prevent misdiagnosis and mismanagement as it never metastasises and responds to local excision.⁹

Learning points

- ▶ Cellular schwannomas, a pseudosarcomatous entity, are rare benign tumours of peripheral nerves.
- ▶ Cellular schwannoma can present clinically and histologically as malignancy, for example, malignant peripheral nerve sheath tumour, synovial sarcoma, fibrosarcoma, etc.
- ▶ Certain histological as well as immunological features are inherent to cellular schwannoma so as to differentiate it from malignant neoplasms.
- ▶ It shows favourable prognosis with total resection appearing to be the curative treatment, with no metastasis and no adjuvant therapy necessary.

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Competing interests None.

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