

Aggressive Melanotic Schwannoma Of The Lumbar Plexus: A Case Report

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Citation

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Abstract

Melanotic schwannoma is a rare variant of schwannoma which usually arises from nerve roots. It occurs in relatively younger age with no sex predilection. Long-term follow up shows that there are chances of recurrences and metastasis even in absence of histological features suggestive of malignancy. Our case describes a 49 year female with 18 years history of low backache. The MRI showed neurogenic tumour arising from lumbar nerve roots, which was confirmed on histopathologic examination to be a melanotic schwannoma. Unfortunately, the patient died of post-operative bleeding.

INTRODUCTION

Melanotic schwannoma is a rare and distinct variant of schwannoma.¹ It usually affects younger adults of either sex with long history of backache.^{1,2} Melanotic schwannoma has been considered as a benign tumour, but it can recur and progress to malignancy, requiring long term follow-up, specially in patients with multiple tumors and/or Carney's syndrome.³ We are hereby presenting a case of a 49 year old female with a 18 years history of backache.

CASE REPORT

A 49-year-old female presented with history of low backache for last 18 years which had increased in intensity for the past 2 years. There was history of radiation of the pain to the left leg. There was no relief with medical treatment. On examination the patient was of average built. The result of various biochemical and haematological investigations were within normal limits. MRI suggested a possibility of neurogenic tumor arising from lumbar plexus. There was no family history or any component of Carney's complex. At surgery, the tumour was richly vascular and the surgeon thought it to be a vascular tumor. The patient died in the post-operative period due to bleeding.

On gross examination, the tumour was a cystic mass measuring 6x5x4.5 cm. The cut surface showed a well-circumscribed unilocular cyst filled with dark brown material (Fig 1). On microscopic examination, the tumor consisted of polygonal to spindle cells with ill-defined cytoplasmic borders. The nuclei were hyperchromatic, some

had prominent nucleoli, while a few of them showed intranuclear vacuoles. There were focal areas of palisading with cellular and hypocellular areas resembling Antoni A and Antoni B patterns. Finely granular to coarsely clumped brown-black pigment was seen at both intracellular and extracellular location (Fig 2). The pigment was positive for Masson Fontana stain and negative for PAS and Prussian blue and thus confirmed it to be melanin. Areas of necrosis, nuclear atypia or mitotic figures were not seen.

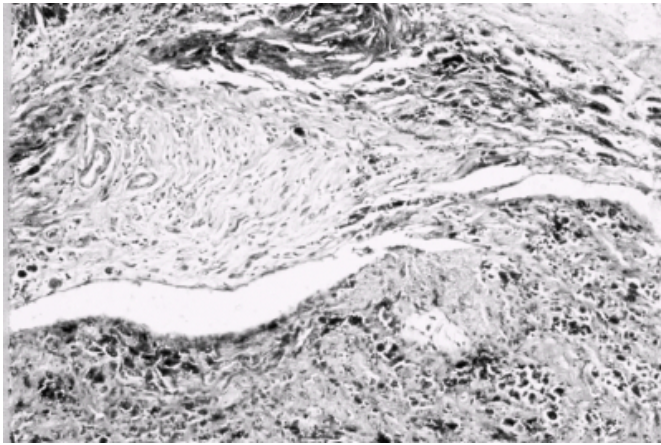
Figure 1

Figure 1: Gross photograph shows encapsulated cystic tumour (6x5x4.5 cm.) containing friable blackish contents.



Figure 2

Figure 2: Photomicrograph shows cellular spindle-shaped areas of schwannoma containing melanin (arrow) (H&E, x550).



DISCUSSION

Melanotic schwannoma is a rare variant of schwannoma.¹ These tumours arise most frequently from the nerve roots resulting in dull aching pain, usually of longer duration and not responding to medical therapy. These patients can have various presentations depending upon the tumour location like sympathetic chain, intramedullary, acoustic nerve, cerebellum, orbit, choroids, soft tissues, oral cavity, heart, oesophageal wall, stomach, bronchus, retroperitoneum, uterine cervix and parotid gland.^{4,5,6,7}

Melanotic schwannoma occur in relatively younger adults (mean 35 years) and have no sex predilection as compared to usual schwannoma, which occur frequently in fifth decade (mean 48 years) with slight female predilection.² Our patient was 49 years old female with low backache of 18 years duration.

One of the largest series of 31 patients with 40 documented melanotic schwannomas had shown other components like spotty pigmentation, endocrine over activity or myxomas in a familial setting labeled as Carney's complex.⁴ These tumours also had psammoma bodies and intra-tumoral adipose tissue, both of which were absent in our case. Hyperparathyroidism has also been reported as a paraneoplastic syndrome in another series.¹

Histogenesis of melanin in schwann cell tumour rests on the fact that neural crest cells migrate and differentiate into divergent tissues like melanocytes, schwann cells, neurons of peripheral nervous system, adrenal medulla, calcitonin producing-C cells of thyroid and mesectodermal cells of the head.⁸ Neural crest is a heterogenous mixture of unipotent,

intermediate potent and totipotent cells and specific differentiation depends on the environment.⁹

These tumours were mostly considered benign because of short follow-up period of most of the cases. The prognosis of all melanotic schwannomas is not good with local recurrence or metastasis in over 40% of the cases. However, histologic criteria for malignancy in melanotic schwannoma are not clearly defined and there is no reliable histopathological indicator of malignant clinical behaviour in melanotic schwannoma.¹⁰ Analysis of total 77 cases with whatever follow-up was available showed that 25 had local recurrence or metastasis, of which 13 died of the disease. Histological features of malignancy were not noted in these fatal cases and mitosis were not high (<2/10hpf).² Sympathetic chain melanotic schwannomas were potentially aggressive. Hence, longer follow up is advisable to look for recurrence or metastasis.

Our patient died in post-operative period due to uncontrolled bleeding. Histologically there were no features to suggest malignancy.

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References

1. Vallat Decouvelaere AV, Wassef M, Lot G, Catala M, Moussalam M, Carvel N, Mikol J. Spinal melanotic schwannoma - a tumour with poor prognosis. *Histopathology* 1999; 35:558-566.
2. Font RL, Truong LD. Melanotic schwannoma of soft tissue. *Am J Surg Pathol* 1984;8:129-138.
3. Goasguen O, Boucher E, Pouit B, Soulard R, Le Charpentier M, Pernot P. melanotic schwannoma, a tumor with a unpredictable prognosis: a case report and review of literature. *Neurochirurgie* 2003; 49: 31-38.
4. Carney JA. Psammomatous melanotic schwannoma. *Am J Surg Pathol* 1990;14:206-222.
5. Jenson OA, Bretlau P. Melanotic schwannoma of the orbit. Immunohistochemical and ultrastructural study of a case and survey of literature. *APMIS* 1990;98:713-723.
6. Terzakis JA, Opher E, Melamed J, Santagada E. Pigmented Melanotic schwannoma of uterine cervix. *Ultrastruct Pathol* 1990;14:357-366.
7. Ranjan A, Chacko G, Chandi SM. Intracerebellar melanotic schwannoma: a case report. *Br J Neurosurg* 1995;9:687-689.
8. Baroffio A, Dupin E, Le Douarin N. Common Precursors for neural and mesectodermal derivatives in the cephalic neural crest. *Development* 1991, 112:301-305.
9. Dupin E, Sextier-Sainte-Claire Deville F, Nataf V, Le Douarin NM. The ontogeny of the neural crest. *CR Acad Sci Paris* 1993;316: 1072-1081.
10. Kuchelmeister K, Lotz C, Schonmayr R, Schachenmayr W. April 2004: Woman in her early fifties with a cervical

intrapinal and extraspinal mass lesion. Brain pathol 2004; 14:453-454.

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