CASE REPORT

Spotty skin pigmentation and multiple blue naevi as cutaneous markers for spinal melanotic schwannoma

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SUMMARY

Spinal melanotic schwannomas are rare spinal tumours with a very poor prognosis in terms of mortality due to difficulty in complete resection and local recurrences. A 67-year-old man presented with acute onset progressive paraparesis for 2 weeks. The patient also had spotty skin pigmentations (blue naevi) in his right lateral thigh. MRI revealed an intradural extramedullary enhancing lesion in the lower thoracic cord level. With a preoperative diagnosis of simple nerve sheath tumour excision was planned. At laminotomy, an infiltrating "en plaque" like lesion reaching up to mid and upper thoracic cord level was identified and excision was carried out. Postoperative histopathology was unique with the identification of melanin and presence of epithelioid cells, with the additional detection of psammoma bodies and adipose-like cells. Melanotic schwannomas though rare and carry poor prognosis must be borne in mind in patients with spotty skin pigmentation and acute onset limb weakness.

Figure 1 Spotty pigmentations (lentigenes) (A) and plaque blue naevi (B) in the upper lateral aspect of the right thigh.

BACKGROUND

Melanotic schwannomas are soft tissue neoplasms accounting for less than 1% of peripheral nerve sheath tumours. The most common sites of the tumour are posterior spinal nerve roots and are described as a part of the Carney complex.¹

CASE PRESENTATION

A 67-year-old man presented with acute onset progressive weakness of both lower limbs of 2 weeks duration associated with difficulty in micturition and constipation. He was a known case of diabetes mellitus and hypertension, on regular treatment. On neurological examination he had reduced tone in bilateral lower limbs, with exaggerated deep tendon reflexes with a power of 3/5. Sensations were diminished from D10 dermatome and bilateral plantar reflexes were extensor. Cutaneous examination revealed multiple 2–3 mm pale-to-dark brown round to irregular macules suggestive of lentigenes over the trunk giving the appearance of spotty pigmentation. We also observed a blue-grey plaque measuring 15×8 cm composed of multiple 2–5 mm blue and dark blue macules and papules over the upper lateral aspect of the right thigh clinically suggestive of plaque blue naevi (figure 1).

INVESTIGATIONS

MRI whole spine revealed an ill-defined abnormal signal intensity lesion measuring 6.1×1.9 cm extending from D8–D12 levels, appearing slightly hyperintense to the cord on T1-weighted sequences (figure 2A) and heterogeneously isointense to hypointense to the cord on T2-weighted sequences (figure 2B). Post gadolinium T1 fat sat sagittal sequences showed avid enhancement of the intradural extramedullary lesion at D8–D12 level (figure 3). There was enhancement in the posterior intradural space extending cranially up to D4 and caudally up to L1. Histopathological confirmation of the cutaneous lesions was not possible as the patient did not consent for skin biopsy since the skin lesions were present since childhood.

DIFFERENTIAL DIAGNOSIS

In view of the association of a spinal tumour with spotty pigmentation (multiple lentigenes) and multiple blue naevi we considered the possibility of Carney complex and evaluated the patient for other associations like cardiac myxoma and endocrine abnormalities which were absent.

TREATMENT

With a preoperative diagnosis of simple nerve sheath tumour, excision was planned. At laminotomy, a black solid tumour was noted in the intradural extramedullary compartment around D8–D12, infiltrating

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Figure 2 (A) T1-weighted sagittal sequence showing intradural extramedullary abnormal signal intensity lesion slightly hyperintense to the cord extending from D8 to D12 level (arrows). (B) T2-weighted sagittal sequence showing the lesion to be heterogeneously isointense to hypointense to the cord (arrows).

the posterior intradural space in an “en plaque” like fashion. Hence a complete resection was not feasible.

OUTCOME AND FOLLOW-UP

Postoperative histopathology was suggestive of psammomatous melanotic schwannoma showing the presence of plump spindle to epithelioid cells arranged in sheets laden with melanin pigment, psammoma bodies and adipose-like cells (figure 4). He

Figure 3 Post gadolinium enhanced T1-weighted fat sat sequences showing avid enhancement of the lesion. There is enhancement of the posterior dural space extending cranially up to the D4 level and caudally up to the L1 level (arrows).

received adjuvant external beam radiotherapy to the spine and is currently on regular follow-up for the last 1 year. His lower limb weakness has been improving with physiotherapy.

DISCUSSION

Spinal melanotic schwannomas (SMS) are rare tumours of Schwann cell origin characterised by cytoplasmic deposition of melanin. They carry a poor prognosis with respect to morbidity and mortality. The most common sites of melanotic

Figure 4 Histopathology of the tumour showing plump spindle to epithelioid cells (A) arranged in sheets laden with melanin pigment (B), psammoma bodies (C) and adipose-like cells (D).

schwannomas are the posterior nerve roots.² They also occur in the sympathetic chain, acoustic nerve, cerebellum, orbit, choroid, soft tissue, heart, oral cavity, oesophageal wall, stomach, bronchus, retroperitoneum, uterine cervix and parotid.³ SMS are symptomatic in two-thirds of cases. Grossly SMS are black, brown or blue, encapsulated and solid or spongy. Microscopically they are incompletely encapsulated with spindle and epithelioid cells, melanin, psammoma bodies and fat cells. These tumours stain positive for S-100 and vimentin and are negative for glial fibrillary acidic protein, actin and keratin.^{4–6}

Majority of the psammomatous schwannomas are associated with Carney's complex. This complex consists of myxomas in the heart, breast, uterus, skin lentiginos, endocrinopathy (Cushing's syndrome due to pigmented adrenal cortical hyperplasia), growth hormone producing pituitary adenoma, epithelioid blue naevi, Sertoli cell tumours of the testis, tumours of the thyroid and ductal adenomas of the breast.^{1 3 4} Our patient fulfilled three of the diagnostic criteria of Carney complex namely histopathologically confirmed psammomatous melanotic schwannoma, spotty skin pigmentation and multiple blue naevi.⁷ However, the blue naevi could not be confirmed by histopathology and the lentigenes were predominantly observed in the trunk.

Most SMS are benign; however, 10% of these tumours are malignant with metastases.^{8–10} Though surgery constitutes the mainstay of treatment, complete resection of the tumour is not always possible, owing to local infiltration and which would need adjuvant postoperative radiotherapy and chemotherapy to

prevent local recurrences and metastases. Preoperative diagnosis is mainly based on MRI. While conventional schwannomas are hypointense on T1-weighted and hyperintense on T2-weighted sequences, melanotic schwannomas are hyperintense on T1-weighted and hypointense on T2-weighted sequences.^{11 12} Appropriate long-term follow-up is required for all SMS, as it may recur or metastasize after more than 5 years, even in the absence of overt malignant histological features.^{2 13}

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Competing interests None.

Patient consent Obtained.

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Learning points

- Melanotic schwannomas are rare intradural extramedullary spinal tumours of the Schwann cell origin which can be mistaken for a simple nerve sheath tumour.
- Spotty skin pigmentation and multiple blue naevi in association with spinal tumour should be considered pointers towards Carney complex, thus suggesting that the spinal tumour would most likely be a psammomatous melanotic schwannoma.

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