Cervical ganglioneuroma: clinical and radiological features of a rare tumour

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Accepted 14 December 2017

DESCRIPTION

Ganglioneuromas are rare benign tumours of neurogenic origin that most frequently develop in the cervical sympathetic chain,^{1,2} with genetic associations demonstrated with Neurofibromatosis type 1 (NF-1). We report the case of a 41-year-old woman with a history of cavum neoplasm at the age of 23. This tumour was classified after assessment of extension as T1N0M0 (tumour, node, metastases). Pathological examination of a biopsy concluded to a keratinising squamous cell carcinoma. The patient was then irradiated (2D Cobalt radiotherapy). The patient presented with gait trouble that appeared a few weeks ago. Clinical examination found a Brown-Séquard syndrome with paresis and loss of proprioception on the left side, and loss of pain and temperature sensation on the right side. Medullary MRI (figures 1 and 2) showed an intradural extramedullary well-circumscribed mass with an hourglass shape from C5 to C6 extending through the left neural foramina. T1-weighted images show

homogeneous hypointense signals, T2-weighted images show heterogeneous hyperintense signals and gadolinium-enhanced images show mildly heterogeneous enhancement. A laminectomy from C4 to C6 was performed with total resection of the tumour. Postoperative course was marked by a complete recovery of the patient. Pathological examination concluded to a ganglioneuroma. Imaging provides valuable information on the size, location, composition of the mass and its relationship to adjacent significant structures, which is helpful in determining a surgical plan. On CT scan,¹ ganglioneuromas are homogeneous, well-circumscribed masses with heterogeneous enhancement after contrast, and calcifications are seen in about 30% of cases. CT imaging may reveal punctate calcifications, which are missing in neuroblastomas.

On MRI, the differential diagnosis of an enhancing dumbbell-shaped mass in the spinal canal extending through the neural foramina includes neurogenic tumours, of which neurofibromas are the most common. Although rare,¹ spinal ganglioneuromas need to be considered for any large mass occupying both the intraspinal and extraspinal compartments, even out of the context of neurofibromatosis. Total surgical resection is the treatment of choice for ganglioneuroma. Surgical decompression of the spinal cord must be undertaken as soon as possible.² Subtotal resection is feasible when total excision is deemed risky.³ Some authors attest that ganglioneuromas may be associated with previous

Figure 1 MRI on T2-weighted sequences in sagittal section shows an hyperintense mass at the levels of C4 and C5 with hourglass appearance.

Figure 2 MRI on T1-weighted sequences in axial section reveals a left lateralised intradural extramedullary mass with intense enhancement after injection of gadolinium, with an extracanal extension.

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To cite: Badri M, Gader G, Bahri K, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-223412

Images in...

radiotherapy. In this case, they appear after an average time of 18 years after irradiation.

Further therapy, chemotherapy or radiotherapy, is not usually required, even for cases with partial excision.

Learning points

- Diagnosis of ganglioneuromas must be discussed at all cervical medullar masses despite of its rarity.
- An enhancing dumbbell-shaped mass in the spinal canal extending through the neural foramina has to lead to think about ganglioneuroma.

Contributors MB and GG: wrote the manuscript. KB: did the bibliographic research and corrected the manuscript. IZ: corrected the manuscript.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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