Primary intradural extramedullary ependymoma: report of two cases and review of the literature

Pierwotny wyściółczak wewnątrztwardówkowy i zewnątrzrdzeniowy: opis dwóch przypadków i przegląd piśmiennictwa

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Abstract

Primary intradural extramedullary ependymomas are very rare. They are called primary in this location as they do not have any connection with the central nervous system. They usually develop from an ectopic ependymal cell nest. To the best of our knowledge, only 10 cases have been described in the literature. We report two cases of large sausage-shaped intradural extramedullary ependymoma in the lumbosacral area.

Key words: primary, intradural, extramedullary, ependymoma.

Streszczenie

Pierwotny wyściółczak wewnątrztwardówkowy i zewnątrzrdzeniowy jest dużą rzadkością. Określenie „pierwotny" odnosi się do braku łączności tych guzów z ośrodkowym układem nerwowym. Guzy te wywodzą się zwykle z ektopowych gniazd komórek wyściółki. Według wiedzy autorów opisano dotąd jedynie 10 takich przypadków. W pracy przedstawiono opis dwóch przypadków dużych, kiełbaskowatych wyściółczaków wewnątrztwardówkowych i zewnątrzrdzeniowych w części lędźwiowo-krzyżowej kanału kręgowego.

Słowa kluczowe: pierwotny, wewnątrztwardówkowy, zewnątrzrdzeniowy, wyściółczak.

Introduction

Tumours of the spinal cord are unusual. In a general hospital, only 5% of spinal tumours are intramedullary, 40% are intradural extramedullary, and 55% are extradural [1]. Ependymoma typically has an intramedullary location and represents 60% of intramedullary tumours [1]. Ependymoma is a glial tumour known to arise from the central nervous system [1]. Primary intradural extramedullary ependymomas of the spinal cord are rare; only 10 cases have been described in the literature. They are called primary tumours in this location as they do not have any connection with the central nervous system [2]. We describe two large sausage-shaped intradural extramedullary ependymomas in the lumbosacral area, the clinical presentations, surgical treatment, and a physiopathological hypothesis of this localization on the basis of the results of the present study and review of the literature.

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Case reports

Case 1

A 24-year-old female was admitted with complaints of lower back pain for the last 3 years, bilateral lower limb numbness for the last 3 months, inability to move both lower limbs and being bedridden for the last 20 days. There was no bowel or bladder disturbance, and no history of trauma to the spine or fever in the recent past.

Her general physical examinations were normal. Her neurological examinations revealed grade 1 to 2 power in both lower limbs, and 50% loss of sensation to all modalities below T12 bilaterally. Deep tendon reflexes and plantar responses were absent bilaterally.

Magnetic resonance imaging (MRI) of the lumbosacral spine revealed a large intradural extramedullary sausage-shaped mass, measuring 17.6 cm (cranio-caudally) × 1.7 cm (antero-posteriorly) × 2.1 cm (transversely), extending from the T12 to S1 vertebral level. The mass was isointense on T1-weighted image, isointense with scattered hyperintense foci on T2-weighted image (Fig. 1A), and post-contrast study showed intense enhancement of the mass (Fig. 1B).

There was no obvious calcification or haemorrhage but it was causing expansion of the spinal canal. The filum terminale and the nerve roots of the cauda equina were not visualized separately. A laminoplasty from T11 to S2 was done. On opening the dura, a large sausage-shaped intradural extramedullary mass was seen extending from T12 to S1. The tumour was capsulated and could easily be separated from the conus and the nerve roots of the cauda equina. The roots and the filum appeared compressed and displaced laterally by the tumour. There was no invasion of filum terminale. It was a pale, pinkish red, highly vascular, firm mass. The tumour was excised totally and watertight dural closure was done. Immediately after the surgery, the patient experienced relief from her back pain, numbness disappeared and motor power in the lower limbs also showed improvement. By the time of discharge, the patient started walking with support. On a follow-up visit after 3 months she came walking without any support.

Case 2

A 21-year-old female was admitted with complaints of lower back pain for the last 1 year and pain radiating to both lower limbs for the last 3 months. There was no weakness, numbness in limbs or bowel and bladder disturbance. There was no history of trauma to the spine or fever in the recent past.

Her general physical examinations were normal. Her neurological examinations were also normal. The straight leg raising test was positive bilaterally at < 60 degrees.

MRI of the lumbosacral spine revealed a large intradural extramedullary mass, measuring 7.8 cm (cranio-caudally) × 1.6 cm (antero-posteriorly) × 2 cm (transversely) extending from the L3 to S1 vertebral level. The mass was isointense on T1-weighted image, isointense with scattered hyperintense foci on T2-weighted image (Fig. 2A), and post-contrast study showed intense enhancement of the mass (Fig. 2B). There was no obvious calcification or haemorrhage. The filum terminale and the nerve roots of the cauda equina are not visualized separately. She was operated on, and laminoplasty L3-S1 was done. On opening the dura, a large capsulated intradural extramedullary mass was seen extending from L3 to S1. The filum and the roots were compressed and displaced laterally by the tumour without any invasion. It was a pale, brownish grey, vascular, soft to firm mass easily separable from the filum and the nerve roots. The tumour was excised totally and watertight dural closure done. Immediately after the surgery, the patient experienced relief from her radiating pain. By the time of discharge the patient started walking with slight back pain. On a follow-up visit after 3 months she came walking without any pain.

Histopathological examination

In both cases the histopathological description was loosely textured tumour comprising small round to cuboidal cells separated by abundant eosinophilic hyaline, mucinous material in a background with microcystic change, interspersed blood vessels of variable size and few papillary structures, no definite ependymal rosettes, showing mild nuclear pleomorphism, insignificant mitosis and scant clear cytoplasm, consistent with myxopapillary ependymoma.

Discussion

Primary intradural extramedullary ependymomas of the spinal cord are extremely rare, in contrast to intramedullary ependymomas or ependymomas arising from the conus medullaris or filum terminale [2-5]. Occa-