Case Report

INTRAMEDULLARY EPIDERMOID TUMORS: A Report of Two Cases

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ABSTRACT

Spinal epidermoid tumors represent less than 1% of all spinal tumors. The vast majority are subdural and extramedullary. Intramedullary epidermoid tumors are extremely rare. There are 54 intramedullary epidermoid tumors reported in literature.

Epidermoid tumors, though seen rarely, must be considered in differential diagnosis of intramedullary mass lesions and the choise for diagnostic test must be Magnetic Resonance Images (MRI).

We report two cases of intramedullary epidermoid tumor located in lumbar and thoracic region with the review of the literature.

Key Words: Epidermoid tumor, Intramedullary tumor, Magnetic resonance imaging, Spinal tumors.

INTRODUCTION

Epidermoid tumors represent 1% of all spinal tumors(1). Most of the epidermoid tumors are subdural or extramedullary. Intramedullary epidermoid tumors are extremely rare (2). Although most epidermoids occur in the lumbar region, intramedullary epidermoid tumors occur most frequently in the thoracicc region (3,4).

We report two cases of intramedullary epidermoid tumors located in lumbar and thoracic region with the review of the literature.

CASE REPORTS

Case 1: A 40-year-old, male patient was admitted with complaints of pain, numbness and weakness in both legs with urinary and faecal incontinence. Numbness first began 1 year ago and with the addition of weakness 6 months ago complaints got worse. Neurological examination revealed paraparesis (motor strength 2/5 right, 3/5 left), hypoesthesia below T1 level, spasticity, babinski +/+, clonus +/+. Plain radiograms of the spine were normal. MRI findings showed intramedullary fusiform dilatation with 3 cm length and 1 cm width, hypointense in T1WI, hyperintense in T2WI, centrally contrast enhancing with nonenhanced hypodense cystic areas after Gadolinium administration (Fig.1). At surgery, T1-T3 laminectomy and total tumor excision were performed. The pathological diagnosis was reported as epidermoid tumor. Postoperative course was uneventful and the patient almost completely recovered from the recorded neurological deficits progressively in time.

Case 2: A 33-year-old female patient was admitted with complaints of numbness in both legs. At the end of the 1st trimester of her pregnancy (3rd month) she began complaining of numbness in legs and the symptom got worse in time until she was admitted to our out-patient clinic at the end of her pregnancy. Neurological examination revealed minimal motor weakness in both lower extremity with bilateral hypoesthesia below L1 level. Plain radiograms of the spine were normal. MRI findings revealed intramedullary mass lesion in between L1-4 levels, hypointense in T1WI and hyperintensie in T2WI, partly contrast enhancing after Gadolinium injection (Fig.2).

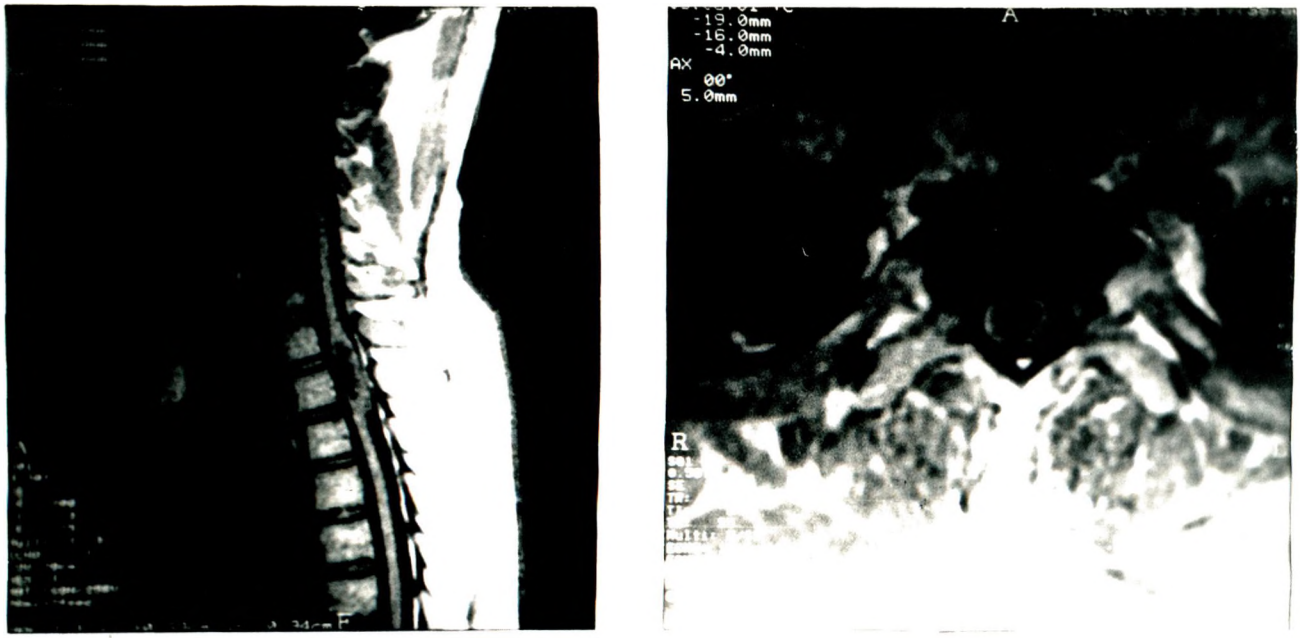

Fig.1.: Sagittal and axial MRI of the thoracic spine demonstrates a well defined mass, hypointense in T1WI, hyperintense in T2WI.

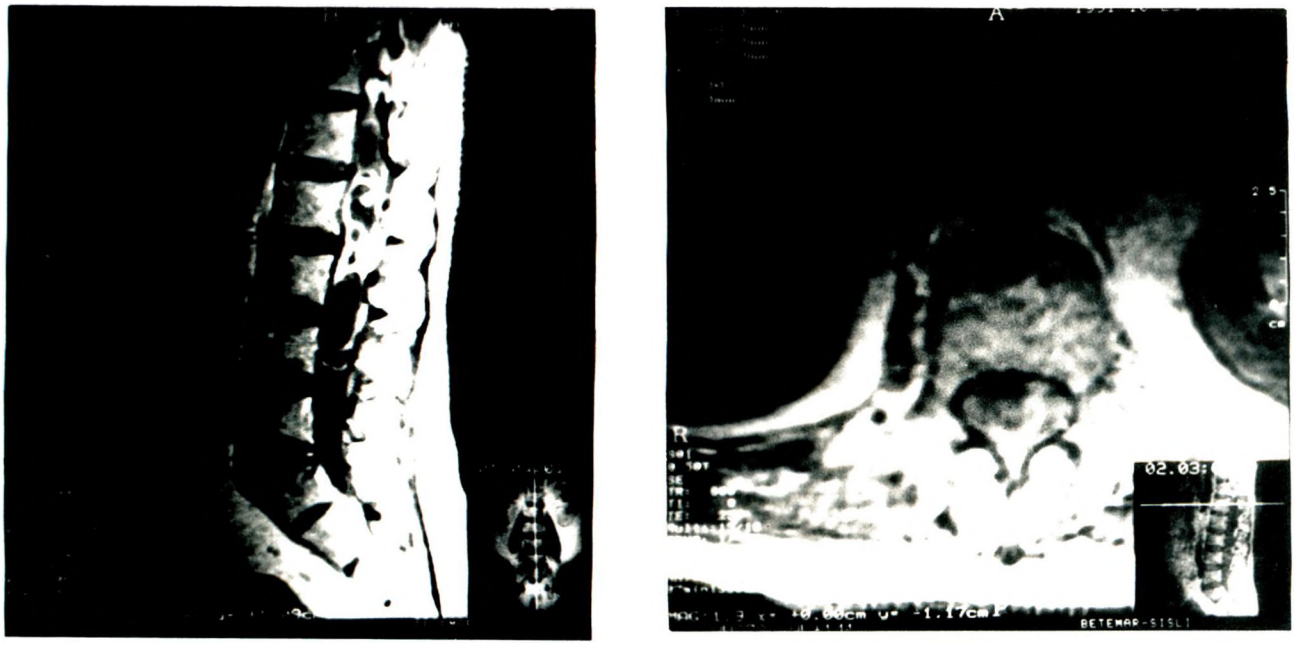

Fig.2.: Sagittal and axial MRI of the lumbosacral spine demonstrates a well defined mass hypointense in T1WI, hyperintense in T2WI, centrally contrast enhancing with nonenhanced hypodense cystic areas after Gadolinium administration.

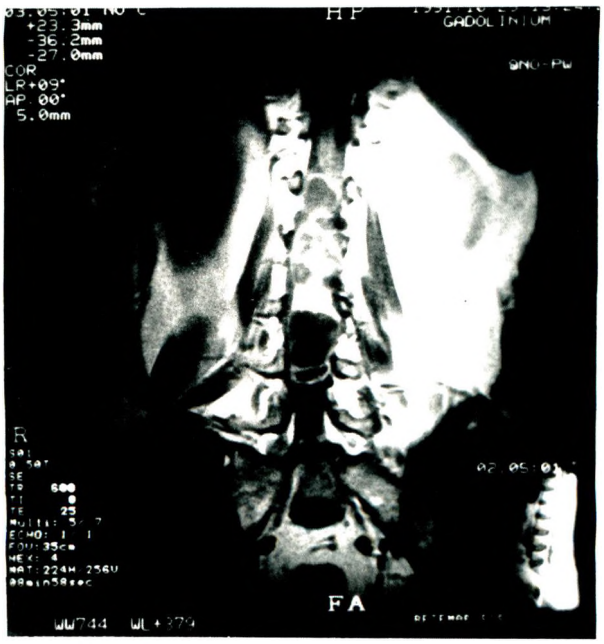

Fig.2.: Coronal MRI of the lumbosacral spine demonstrates a well defined mass hypointense in T1WI, hyperintense in T2WI, centrally contrast enhancing with nonenhanced hypodense cystic areas after Gadolinium administration.

At surgery, L1-L3 laminectomy and total tumor excision were performed. Pathological diagnosis was reported as epidermoid tumor. Postoperative course was uneventful and patient was discharged without deficit.

DISCUSSION

Spinal epidermoid tumors represent less than 1% of all spinal tumors (1,4). The vast majority are subdural and extramedullary. Intramedullary epidermoid tumors were extremely rare (4,5). The first intramedullary epidermoid tumor was reported by Chiari in 1883 (6). The first successful surgical treatment of an intramedullary epidermoid tumor was reported by Gross in 1934 (7). There are 54 intramedullary epidermoid tumors reported in literature (1). The review of the 54 intramedullary epidermoid tumors showed a male: female ratio of 1.14:1. (29 male and 25 female). The patients ranged in age from 2 to 71 years, with an average age of 32 years. These tumors are well tolerated; duration of symptoms varied from 2 days to 53 years, with an average of 5 years. Although most epidermoids occur in the lumbar region, intramedullary epidermoid tumors occur predominantly in the thoracic region; they may, however, occur anywhere in the spinal cord (3). Localizations of tumors in our cases (one in the thoracic and the other in lumbar region) correlate well with the literature.

Intramedullary epidermoid tumors show the classical signs and symptoms of spinal cord tumors. The usual initial complaints are pain and paraparesis (4). Both of our cases had complaints of weakness in legs at admittance.

Epidermoid tumors are considered to be congenital. They are related to the inclusion of the ectoderm without dermal elements, at the time of the closure of the neural tube, between the third and fourth weeks of fetal life (1,3,4,8). They are usually assocaiated with other congenital spinal anomalies (1,2,7,8-10). In some cases, however, the more caudally localized spinal cysts seem to be acquired after single or multiple lumbar spinal punctures (4-9). Also a history of trauma, in the aetiology of epidermoids has been reported by several authors (4).

Macroscopic appearance of epidermoid tumors are typical, Cruveilhier named them as "pearly tumors"(11). Epidermoid tumors are lined by simple stratified squamous epithelium resting on an outer layer of connective tissue. It is filled with a soft and waxy material rich in cholesterol crystals, but contains no skin appendages (4,8-11).

The diagnosis of intramedullary lesions before surgery is diffcult. Plain X-rays and myelography show signs of an expanding lesion (2,4,12). CT shows homogeneous hypodense mass not enhanced by contrast material (2,4,12). MRI is the best diagnostic imaging technigue for intramedullary epidermoid tumors. In MRI, epidormoids are hypodense on T1 and may show some peripheral enhancement with Gadolinium administration, whereas T2-weighted images of these lesions show a hyperdense mass (2,4,13). MRI findings of our cases correlate well with the literature.

Differential diagnosis includes other intramedullary mass lesions such as dermoid tumor, glial tumors, hydromyelia, cysts and abscess (4).

Surgical management of epidermoids is total excision, usually with a good surgical outcome (1,2,5,9). Since it is known that epidermoid cyst's content produces aseptic meningitis at surgery, protection against widespread subarachnoid dissemination of the cyst is necessary (4). When the capsule of the intramedullary epidermoid cyst is thick and adherent to the surrounding cord tissue, total resection is impossible. Even subtotal resection, however, will arrest progression of neurological deficits for many years (2,9,10,13). If the symptoms recur, reoperation is indicated (13). Tumors of both of our cases have totally been removed and they had become symptom free in their follow ups.

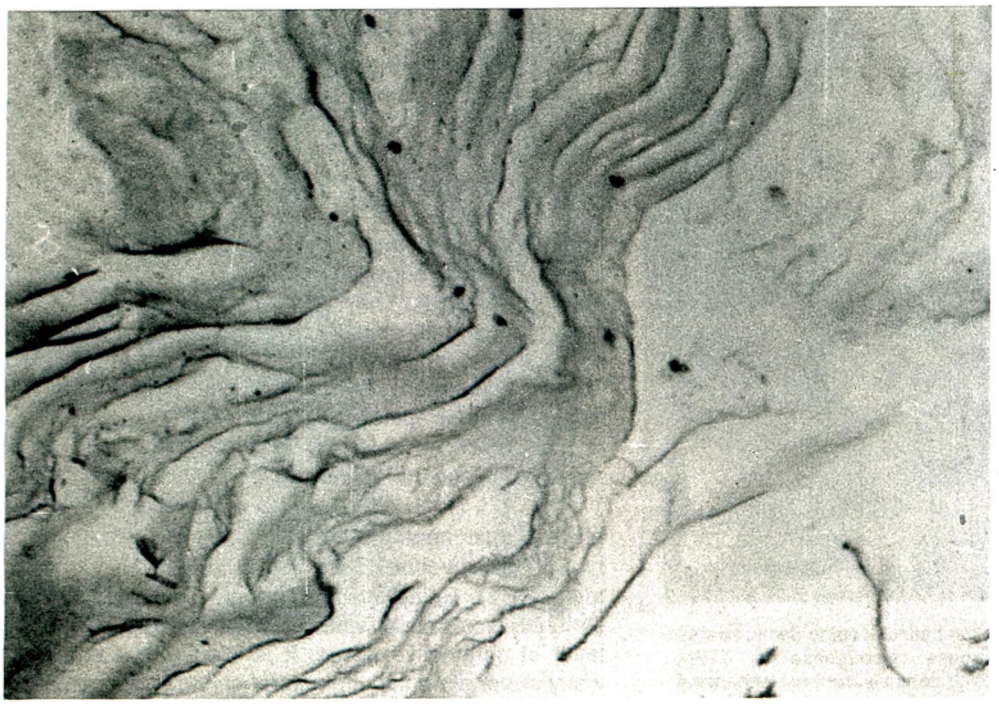

Fig.3.:
Histological features of the cyst, containing skin and skin appendages (H&E x 100).

As a result epidermoid tumors though seen rarely must be considered in differential diagnosis of intramedullary mass lesions and the choice for diagnostic test must be MRI.

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