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Intramedullary Dorsal Schwannoma in an Adolescent: A Case Report

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Abstract

Intramedullary tumors are rare among primary tumors of the central nervous system. They most often affect young adults, with no gender predominance. Spinal schwannomas account for approximately 30% of all spinal tumors. The most common location is in the thoracic region, and they are most often intradural. The patient was a 16-year-old adolescent with no known medical or surgical history, who was admitted for lumbodorsalgia. Examination revealed paraplegia and sphincter disorders. MRI of the thoracolumbar spine revealed an intramedullary tumor extending from D1 to D5. The patient underwent tumor resection followed by functional motor and sphincter rehabilitation, with an unfavorable clinical outcome apart from a slight recovery of sensitivity. Immunohistochemical analysis of the surgical specimen confirmed the diagnosis of schwannoma. The discovery of an intramedullary schwannoma is not unusual. The diagnosis was confirmed by histology, and treatment was surgical.

Keywords

Intramedullary Tumor, Surgical Resection, Schwannoma

1. Introduction

Spinal schwannomas account for approximately 30% of all spinal tumors [1]. These tumors are generally benign. The nonspecific clinical picture usually combines spinal or radicular pain with sensory-motor deficits and/or sphincter disorders that appear gradually or subacutely [2]-[4].

Medical imaging, in this case magnetic resonance imaging (MRI), contributes significantly to the early positive diagnosis of these tumors. The most common location was in the thoracic region (40%), and 62% of schwannomas are intradural

[3]. Pathology provides certainty regarding the diagnosis and histological type of the tumor. The treatment of choice for most of these intramedullary tumors is surgical resection.

We report here the histological profile and progression of a patient who underwent surgery in our department.

2. Case Report

The patient was a 16-year-old girl with no known medical or surgical history, admitted for functional impotence of the lower limbs associated with paresthesia of the lower limbs, characterized by tingling and pins and needles, with symptoms beginning approximately three (3) months earlier. The clinical examination revealed: fairly good general condition, preserved consciousness with a Glasgow score of 15/15, spinal syndrome: dorsolumbar back pain, lesional syndrome: paresthesia of the lower limbs with hypoesthesia extending up to D4, sublesional syndrome: flaccid-spastic paraplegia with motor strength rated at 1/5, osteotendinous reflexes were lively, polykinetic, and diffuse in the lower limbs. Existing sphincter disorders include anal incontinence and acute urinary retention, requiring the use of a urinary catheter for more than 60 days.

The MRI of the thoracolumbar spine revealed a tumor-like intramedullary lesion extending from the D1 to the D5 vertebrae (Figure 1).

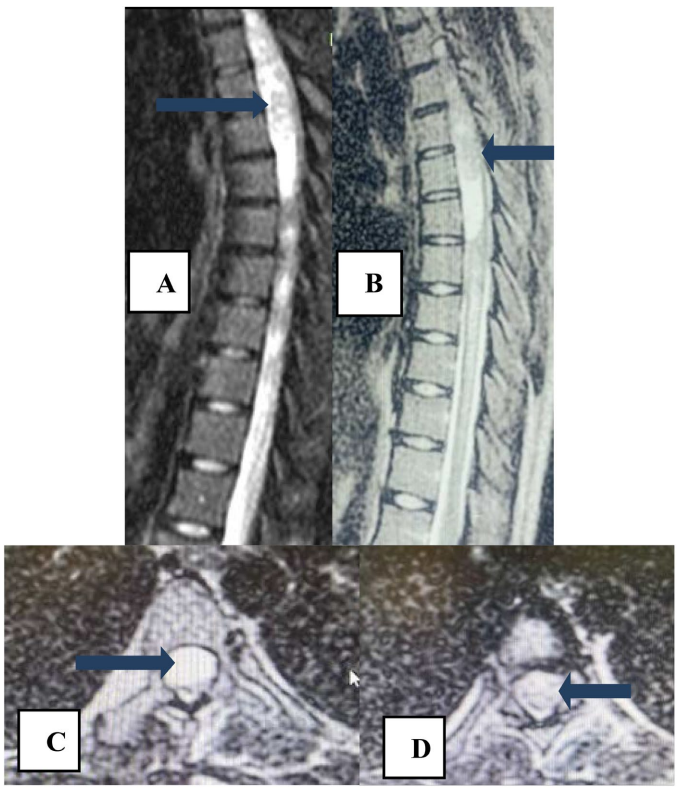

Figure 1. MRI of the thoracic spine, sagittal slices, (A, C and D) T2 sequence and (B) T1 sequence showing a heterogeneous intramedullary lesion with a fleshy and cystic appearance (C) and irregular contours, consistent with a tumor (D).

Given the clinical picture of upper spinal cord compression and the evidence of spinal cord injury at D1 - D5 on MRI, surgery was indicated and performed. Tumor resection was performed to obtain a definitive histological diagnosis.

The surgical technique consisted of a laminectomy from D1 to D5, opening and fixation of the dura mater, followed by opening of the arachnoid membrane under a microscope. A large, tense spinal cord was discovered, and a median myelotomy was performed with careful dissection of the tumor. Complete en bloc resection of a slightly indurated, yellowish lesion was achieved. Preservation of the root carrying the tumor could not be guaranteed because the root had not been identified.

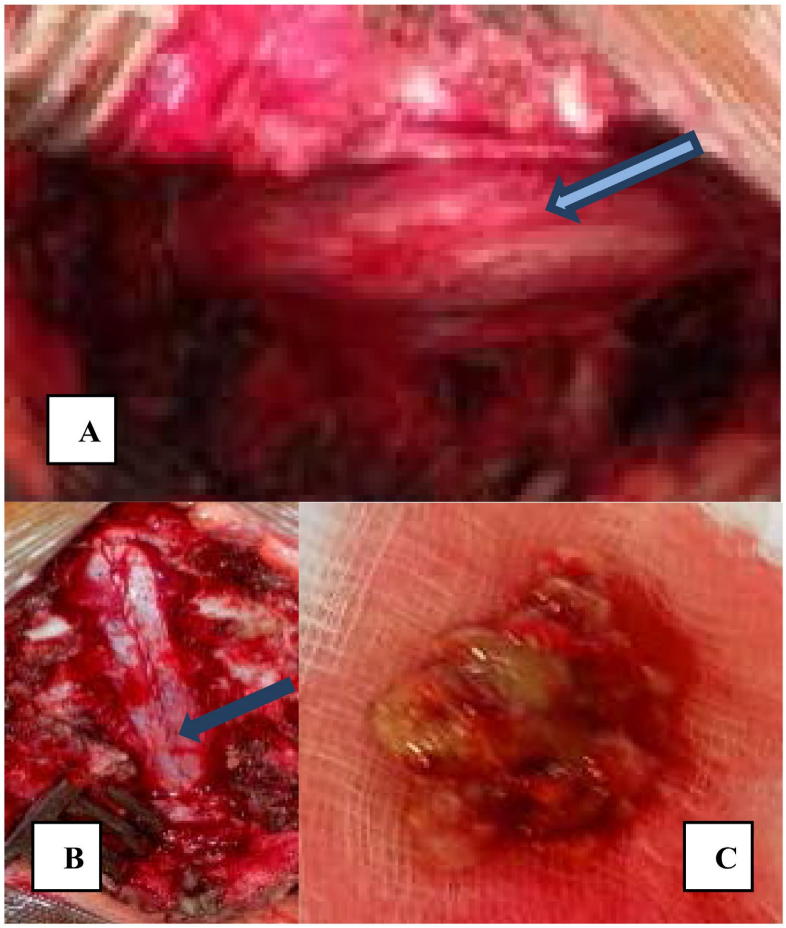

Figure 2. Intraoperative images. (A) Laminectomy D3 - D5 and opening-fixation of the dura mater revealing a large dorsal spinal cord. (B) The remainder of the spinal cord after excision and dural closure. (C) Surgical specimen from intramedullary tumorectomy.

After eighteen (18) months, there was a slight improvement followed by stagnation in motor strength, rated at 2/5 in the lower limbs, an improvement in sensitivity, and persistent sphincter disorders. The pathological examination of the surgical specimen and, above all, the immunohistochemical study of the tumor were consistent with the diagnosis of a schwannoma (see **Figure 2**, **Figure 3**).

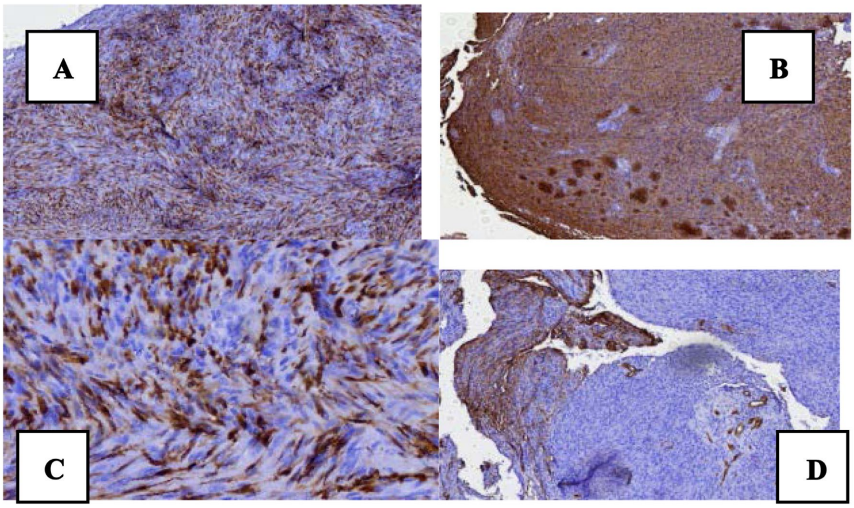

Figure 3. Immunohistochemical study of the surgical specimen of the intramedullary lesion consistent with the diagnosis of schwannoma. (A) Calretinin antibody × 100, (B) PS100 antibody × 100 strongly positive, (C) Calretinin antibody × 400, (D) CD34 antibody × 100.

3. Discussion

Kernohan was recognized as the first neurosurgeon to report a case of intramedullary schwannoma in 1952, although Penfield had already described a similar intramedullary lesion with schwannoma characteristics in 1932 [5].

Schwannomas originate from Schwann cells, which are normally absent from the central nervous system, which may explain the rarity of intramedullary schwannomas.

The age of onset of the tumor in our case was adolescence (16 years old). This young age corroborates certain data in the literature [1].

The revealing symptoms in this specific case are dorsolumbar pain, flaccid-spastic paraplegia, and D4 hypoesthesia associated with sphincter disorders. This is similar to the various clinical pictures described in the literature, with signs of slow spinal cord compression in the foreground [1] [6].

The diagnosis was delayed, with an average delay between the onset of symptoms and the first specialist consultation of 3 months (90 days). Our delay is greater than that reported by Zabsonre DS [6], who found a delay of 79 days, but much less than that reported by Hamdane MM [7], who found a delay of 9.25 months.

In our practice in Chad, socioeconomic realities and ignorance are very often the cause of late consultations, with alternative therapies being used most often at first. This could explain the late consultation on the one hand, and on the other hand, the gradual and insidious onset of slow spinal cord compression syndrome could justify this delay.

On imaging (MRI), the tumor is located in the dorsal region. The dorsal location was reported by several authors [2] [8]. Neuroimaging remains the key examination for establishing the topographical and lesion diagnosis and guiding the

therapeutic approach for intramedullary tumors [9].

Surgery is the treatment of choice for spinal schwannomas, the aim of which is to relieve radicular and/or medullary compression, preserve the anatomical and functional continuity of the affected nerve as far as possible, and perform a complete surgical resection. In our case, tumor excision was possible, unlike in the study conducted by Raghavendra N. [10], in which separation of the tumor from the spinal cord was impossible, leading to the choice of radiotherapy. The choice of posterior approach is guided by the location of the tumor, the level of the lesion, and its intra- and extradural relationships [6] [8] [9].

Immunohistochemistry was essential for confirming the diagnosis in the absence of neurofibromatosis in our case. The positive diagnosis was clearly established by the strong positivity of the S-100 marker in favor of schwannoma, distinguishing it from ependymoma. Calretinin is also more specific to schwannoma, whereas CD-34, although positive in schwannomas, is also positive in neurofibromas. Preliminary histology revealed a benign encapsulated tumor developed at the expense of Schwann cells [9]-[11].

Motor physiotherapy sessions following surgical treatment were indicated for paraplegia. This is an important part of the treatment of neurological deficits, improving the outcome of surgical treatment and increasing the patient's chances of neurological recovery.

After eighteen (18) months of follow-up, there was a slight improvement with motor strength remaining at 2/5 in the lower limbs and an improvement in superficial sensitivity, but sphincter disorders persisted. This can be explained by spinal cord compression lasting more than three months, with definite spinal cord lesions limiting the chances of complete recovery. This has been reported by most of the authors cited above. However, ongoing maintenance physiotherapy is necessary as part of the ongoing treatment to prevent muscle atrophy in the lower limbs.

4. Conclusion

Spinal schwannomas are relatively uncommon among all spinal tumors. They are most common in young people. The clinical presentation is that of nerve or spinal cord compression, often with gradual onset. A definitive diagnosis is made by histology. Management is multidisciplinary.

Conflicts of Interest

The authors declare no conflicts of interest relating to the completion of this work.

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