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Mature Dorsal Teratoma Revealed By Paraplegia In A Two-Year-Old Child: A Case Report

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Abstract:
Spinal teratomas, particularly in the pediatric population, present a diagnostic and therapeutic challenge due to their rarity and diverse clinical presentations. We present a case of a dorsal mature teratoma in a two-year-old child, a rare occurrence not associated with dysraphism. The patient's clinical history revealed delayed walking followed by progressive paraparesis leading to incomplete paraplegia with trophic disorders in both lower limbs. Diagnostic imaging, particularly MRI, played a crucial role in preoperative assessment, while histopathological examination confirmed the diagnosis. Surgical resection remains the mainstay of treatment, aiming for gross total removal to minimize recurrence risk. In our case, complete excision of the tumor was achieved, leading to partial motor improvement postoperatively. Adjuvant therapy such as radiotherapy is reserved for malignant teratomas, while chemotherapy has limited efficacy. Overall, spinal teratomas require a multidisciplinary approach for diagnosis and management, with careful consideration of individual patient factors to optimize outcomes and minimize morbidity. This rare entity in the pediatric population should be included in the differential diagnosis of extramedullary intradural spinal cord compression, highlighting the importance of early diagnosis and complete surgical resection for favorable outcomes.

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I. Introduction:

Teratomas account for 03% of childhood tumors, with the overall incidence of mature teratomas estimated at 0.15% and 0.18% of all spinal tumors [1,2].

In pediatric patients, 5-10% of spinal lesions are intra spinal teratomas, whereas the incidence in adults is much lower [3]. However, these tumors are thought to occur more frequently in children and infants than in adults [4].

Intradural extra-spinal teratomas are a rare condition, often associated with spinal dysraphism.

By definition, teratomas are tumors containing differentiated tissue from one, two or all three primitive germ layers (endoderm, mesoderm and ectoderm); primitive germ cells can proliferate into benign lesions (mature teratomas and immature grade I teratomas) ; into malignant lesions (high-grade immature teratomas and malignant germ cell tumors) or, exceptionally, into benign lesions with secondary malignancy (cancerized teratomas).

Mature teratomas are benign tumors (albeit with a significant risk of recurrence). Depending on the extent of its immature neuroepithelial component).

II. Materials And Methods:

We report an exceptional case of a dorsal mature teratoma in a two-year-old child, revealed by slow dorsal spinal cord compression, whose clinical history began with delayed walking at the age of one, followed by progressive onset of paraparesis that later worsened to incomplete paraplegia with trophic disorders in both lower limbs.

Observation:

Spinal teratomas are rarely seen and occur as a result of a combination of cells, which originate from the three germinal layers; however, endodermal layers do not always accompany the tumor.

The tumors arising from only two germinal layers are called “teratoid” or “bigerminal teratoma”. Teratomas are also referred to as teratomatous cyst, cystic teratoma, teratoid tumor, or atypical teratoma.(2–4) While the pathogenesis of spinal teratoma remains controversial, there are some theories regarding its development, the most accepted of which is based upon the fact that the primordial germ cells that originate from the yolk sac are located at a different site due to alteration during cell migration in early embryogenesis.(1,4,5)

Our case is a two-year-old child, with a history of hypothyroidism under treatment, presenting with a pyramidal syndrome consisting of incomplete spastic paraplegia, vivid ROTs on both sides, abolished cutaneous-abdominal R.s and Babinski present bilaterally. Sensitivity to all modes was preserved, as was amyotrophy in both lower limbs. However, on physical and radiological examination, we did not find any signs of dysraphism in our patient. Spinal cord MRI revealed an extramedullary cystic expansive process at T3 and T4 with T2 hypersignal, T1 hyposignal, oval peripheral enhancement, well limited, compressing and laminating the opposite dorsal medulla, which was T2 hypersignal. EMG of the 02 lower limbs was unremarkable.

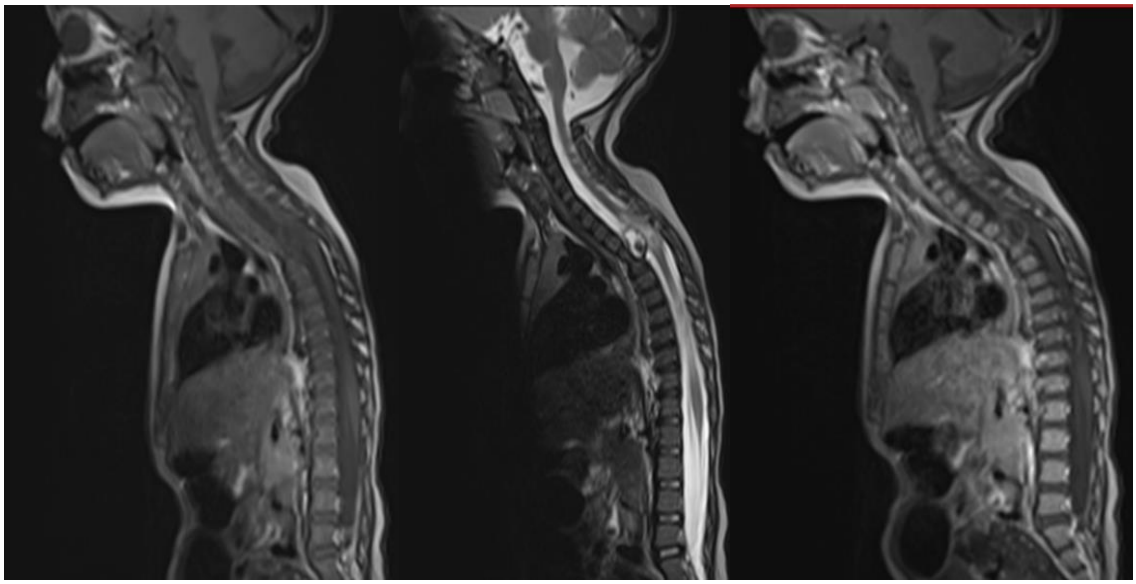

Fig 01: T1, T1 gado, and T2-weighted sagittal section of spinal cord MRI

III. Discussion:

Spinal teratomas, whether intradural or intramedullary, present a diagnostic and therapeutic challenge due to their rarity and varied clinical manifestations.

The first patient was reported to be diagnosed in 1863 by Virchow.(4,6) Intradural spinal teratomas account for up to approximately 0.1% of all the spinal tumors, and they may be extradural, intradural or intramedullary (1,2,6,7). Poeze determined 83 patients with spinal teratoma in 1999, of which 31 (37%) were intramedullary teratoma, and 58% of teratomas were detected in adult patients.(8,9, 10)

Other abnormalities such as diastematomyelia, myelomeningocele and tethered cord may accompany spinal teratoma.(2,3,6,10,11)

Diagnostic imaging, particularly MRI, is crucial for preoperative assessment, although histopatho logical examination remains necessary to confirm the diagnosis (12,13).

Treatment primarily involves surgical resection, aiming for gross total removal whenever feasible to minimize the risk of recurrence (14-16). However, the dense adherence of these tumors to surrounding neural structures may necessitate subtotal resection to avoid neurological deficits. Adjuvant radiotherapy is reserved for malignant teratomas, with no established role for chemotherapy (17).

The diagnosis of spinal teratomas in children and adults relies on a combination of clinical presentation, radiological imaging, and histopathological examination (18). While spinal teratomas are more common in pediatric patients, constituting 5-10% of spinal lesions, they are far less prevalent in adults, accounting for only a small fraction of spinal tumors (17).

In our case the delay in diagnosis is due to:

- neurological examination is difficult in young children, especially in non-ambulatory infants with poor verbal skills, and the neurosurgeon is consulted only as a last resort.
-the insidious nature of certain clinical manifestations, due to the slow evolution of this process.

Other subtle findings on clinical examination are used to reinforce the diagnosis; clinical features such as progressive scoliosis and abnormalities such as skin stigmata and spinal dysraphism are often associated with these cases, and can provide important clues to the diagnosis (18).

MRI serves as the primary diagnostic tool, providing valuable information about tumor location and involvement of adjacent structures (12,13). However, tissue diagnosis through histopathological examination is essential for confirming the diagnosis and determining tumor characteristics.

Surgical resection is the mainstay of treatment for spinal teratomas, with the goal of achieving gross total removal whenever possible (19-23). In pediatric patients, complete removal may be more achievable, with reported success rates of 61.8% for intramedullary teratomas. However, in cases where tumors are densely adherent to critical neural structures, subtotal or partial resection may be considered to minimize the risk of neurological deficits (18,24,25).

Our patient underwent surgery with complete excision of the left anterolateral solid-cystic double-component tumor, highly adherent to the medulla, through a laminotomy extending from D2 to D5. Anatomopathological examination concluded that the tumor was a mature teratoma. Postoperatively, the patient showed partial motor improvement and was referred for rehabilitation.

Symptomatic recurrence rates for mature teratomas are generally low, even following incomplete resections, suggesting that aggressive surgical approaches may not always be necessary.

Adjuvant therapy, such as radiotherapy, is typically reserved for malignant teratomas, whereas benign tumors may not require additional treatment beyond surgery (17). Chemotherapy has not demonstrated efficacy in the management of spinal teratomas and is not commonly used. Intraoperative electrophysiologic monitoring is invaluable during surgical resection, providing real-time feedback to guide the extent of tumor removal and minimize the risk of permanent neurological sequelae (18,26).

Overall, spinal teratomas represent a rare but important subset of spinal tumors that require a multidisciplinary approach to diagnosis and management. While surgical resection remains the cornerstone of treatment, careful consideration of individual patient factors and tumor characteristics is essential to optimize outcomes and minimize morbidity.

IV. Conclusion:

In conclusion, thoracic spinal teratomas are uncommon in the pediatric age group. The tumor is generally located at the conus site in adult patients, while it is frequently larger sized in children and located at thoracic and cervical sites. Should the tumor not be removed radically, the survival without recurrence may be extended

We report a very rare case of dorsal mature teratoma in a two-year-old child, not associated with dysraphism, the particular interest of our case apart from its exceptional location lies in:

-Early diagnosis, which is essential in order to resect the tumor as early and completely as possible.

-Total surgical resection, which is the goal for these tumors due to their benign nature.

-The neurological prognosis depends on the histological nature (benign in 90% of cases) and the patient's initial clinical condition.

This rare or even exceptional entity in the pediatric population should be included in the differential diagnosis of extramedullary intradural spinal cord compression.

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