

Unusual Association of Intra-Dural Lipoma and Myelomeningocele: Case Report and Literature Review

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ABSTRACT

We report the case of an 18-month-old girl who presented a complex spinal dysraphism combining an intra dural lipoma and myelomeningocele. The coexistence of intradural lipoma and myelomeningocele is very rare. We will focus on the contribution of imaging to the diagnosis and follow-up of spinal dysraphism.

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Introduction

Spinal dysraphisms are a heterogeneous group of congenital malformations involving the bony component of vertebrae or spinal cord or both. Spinal dysraphisms include congenital malformations of the spinal column and cord. These result due to disorders during early embryonic stage, 2–6 weeks of gestational age [1].

We report a case of Complex form of spinal dysraphism including intra dural lipoma and myelomeningocele.

More than two types of spinal dysraphism may co-exist in the same patient, which are extremely uncommon and a few of them have been reported in the literature.

Case report

We report the case of a 18-month-old girl who presented on clinical examination with a soft lumbar mass and paresis of the lower limbs.

The child underwent a medullary magnetic resonance imaging (MRI) which revealed a medullary cone in normal position fixed by an intracanal oval intramedullary dorsal mass (extending opposite D9 to D12), hyperintense on T1-weighted images, hyperintense on T2-weighted images, not erased in STIR sequences, with no enhancement after injection, which matches with intra dural lipoma (Figure 1a, b, c).

It revealed also myelomeningeal herniation through a localized bone defect in the posterior arch of the lumbar vertebra (L2), forming a small myelomeningocele (Figure 2 a,b).

After a multidisciplinary meeting, the infant has been operated and both of the masses were removed. In the early postoperative period, there was no complication. Three months after the surgery, the patient improved significantly.

Discussion

Intra dural lipoma is a rare lesion representing 1% of medullary tumors, 4% of spinal lipomas, and can be localized to different levels although the most frequent topography is the lumbosacral level where the attachment to the marrow causes the low position of the medullary cone [2].

An intradural lipoma refers to a lipoma located along the dorsal midline that is contained within the dural sac [3].

Intra dural lipoma is a result of a premature disjunction, before the neural tube closes, between the neuroectoderm and the ectoderm allows the adjacent mesenchyme to come between these two tissues and adhere to the lumen of the neural tube, lined with ependyma primitive. This induces the differentiation of the mesenchyme into fatty tissue [4].

This event takes place after the disjunction between neuroectoderm and ectoderm and for this reason, the lesion is covered by the skin which appears intact [4]. The fatty tissue remains attached to the dorsal surface of the placode in space sub-pial and in contact with the pia mater. Depending on its size, it can repress the pia mater seeming to appear in spaces subarachnoid but the dura remains intact and completely borders the lipoma. It can invaginate in the marrow spinal to the ependymal canal, but pure intramedullary localization is rare. The spinal canal is often enlarged and the lipoma may increase in size with the growth of the child [4].

Myelomeningoceles account for more than 98% of open spinal dysraphisms [5]. In MMC, the protruding neural placode extends beyond the skin surface as there is enlargement of the adjacent subarachnoid space [6]. Herniation of the fluid-filled sac with neural elements points towards MMC, which is an open defect and is seldom referred for imaging. The neural placode in MMC is seen protruding beyond the dysraphic spinal defect due to the subarachnoid cystic component [7].

Magnetic resonance imaging (MRI) is the investigation of choice for evaluation of spinal dysraphism. These disorders can be complicated and variable in imaging appearance and can be confusing for the inexperienced radiologists. A detailed and systematic analysis of each component of the spine is crucial for obtaining a correct diagnosis [8]. Pre-operative MRI allows to obtain anatomic characterization of the different components of the malformation, the relationship

between the placode and nerve roots, presurgical assessment of the malformation; hydromelia, hydrobulbia, Chiari II malformation and hydrocephalus [1].

Evaluation of spinal dysraphism associated with masses on MRI can be confusing and should be achieved by step-by-step evaluation of the contents of these lesions (fat, fluid, soft tissue, and neural components), extent (intraspinal, both intra and extraspinal, subcutaneous), the components passing through the dural and vertebral defect, and interface of the intratumoral fat with the neural placode and with the subcutaneous fat [8].

In our case the clinical features was suggestive of myelomeningocele, the preoperative MRI allows to identify the intradural lipoma and confirmed the lumbar myelomeningocele.

Conclusion

Congenital malformations of the spine and spinal cord can be complex and variable in imaging appearance. MRI is the modality of choice for the preoperative characterization of the different types of spinal dysraphism and in the diagnosis of associated malformations.

The knowledge of the different type these malformation with structured analysis on MRI of the anatomic components eases the diagnosis even for the beginner radiologists.

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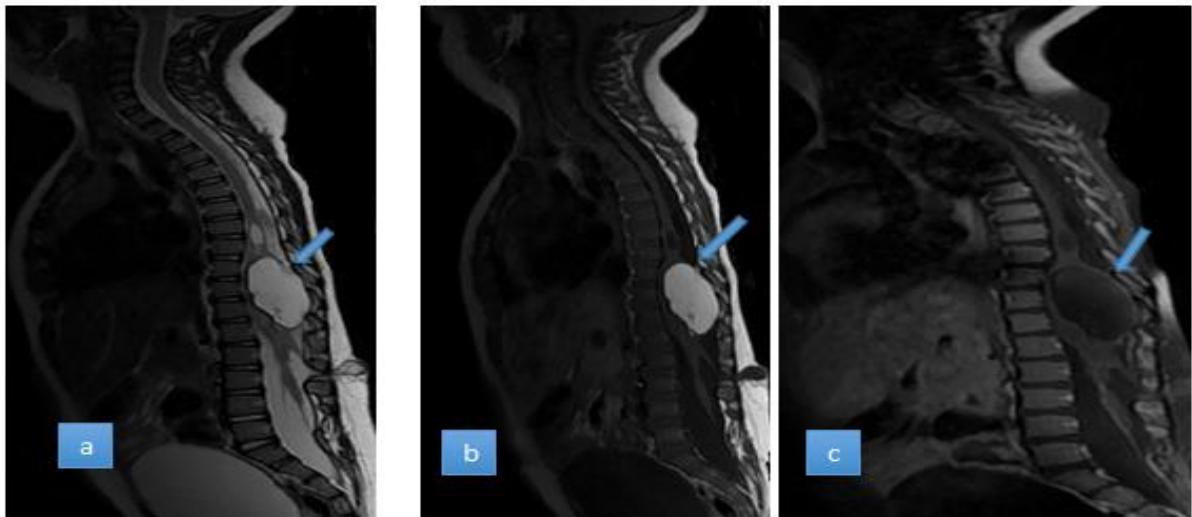


Figure 1 . MRI imaging showing medullary cone in normal position fixed by an intracanal intramedullary dorsal mass : hyperintense on T2-weighted images (a), not erased in STIR sequences (b), with no enhancement after injection (c), which matches with intra dural lipoma.

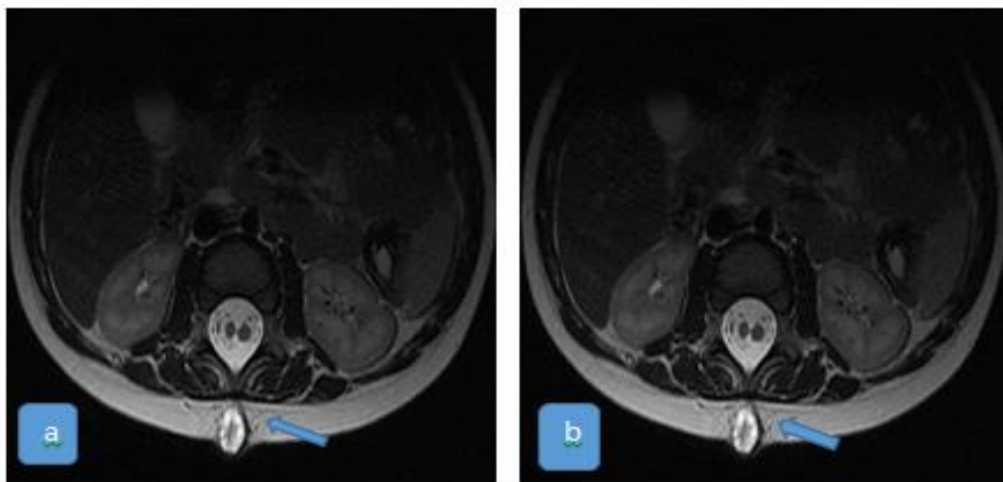


Figure 2. a,b. MRI imaging (T2 images) : myelomeningeal herniation through a localized bone defect in the posterior arch of the lumbar vertebra (L2), forming a small myelomeningocele.