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EXTRAMEDULLARY EPIDERMOID IN A CASE OF MYELOMENINGOCOELE: AN UNUSUAL ASSOCIATION

Neurosurgery

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ABSTRACT

Spinal epidermoid cysts are rare constituting between 0.5% to 1% of all spinal tumors but accounting for up to 10% of intraspinal tumors in children. They are most often seen in the lumbosacral spine. Males are more frequently affected than females. Epidermoid cysts arise from pathologic displacement of epidermal cells into the spinal canal. They can be acquired from trauma, lumbar puncture, surgery such as myelomeningocele closure or arise as congenital lesions, like when there is improper closure of the neural tube, which results in the inclusion of epiblasts in the neural tube. Forty percent are intramedullary and sixty percent are extramedullary. Myelomeningocele is an entity which is common in the lumbosacral region due to congenital neurulation defect. The prevalence of myelomeningocele is approximately 0.8 to 1 per 1000 live births worldwide. The anatomical location may vary as cases of cervical, cervicothoracic, thoracic, thoracolumbar, lumbar and sacral have also been documented. The sac usually contains nerve fibres or neural tissues, however sometimes it may contain intradural lipoma or dermoids. Here we present a rare case of a 1 year 6 month old male child presenting with with dorsolumbar myelomeningocele with symptoms of urinary and bowel incontinence and weakness of both lower limbs found to have extrameduallary epidermoid as a content of myelomeningocoele sac. The concurrence of a congenintal neoplasm within the spinal canal associated with myelomeningocele seems to be a very rare event as very few such cases have been reported worldwide to the best of our knowledge.

KEYWORDS

INTRODUCTION

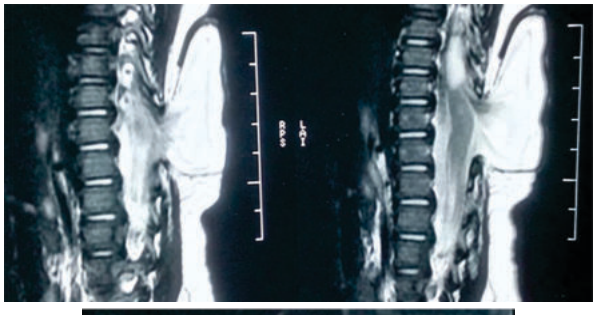
Epidermoid cysts may be described as rare benign neoplasms, which arise from the epidermal cells, within the neuroaxis . In 1835 these were first described by by Cruveilhier as “tumeurs perleés” (pearly tumors). Spinal epidermoid cysts are rare constituting between 0.5% to 1% of all spinal tumors but accounting for upto 10% of intraspinal tumors in children(1). They are most often seen in the lumbosacral spine. Males are more frequently affected than females. Epidermoid cysts arise from pathologic displacement of epidermal cells into the spinal canal. They can be acquired from trauma, lumbar puncture, or surgery such as myelomeningocele closure, with implantation of viable epidermal elements, or arise as congenital lesions, like when there is improper closure of the neural tube, which results in the inclusion of epiblasts in the neural tube. Forty percent are intramedullary and sixty percent are extramedullary.

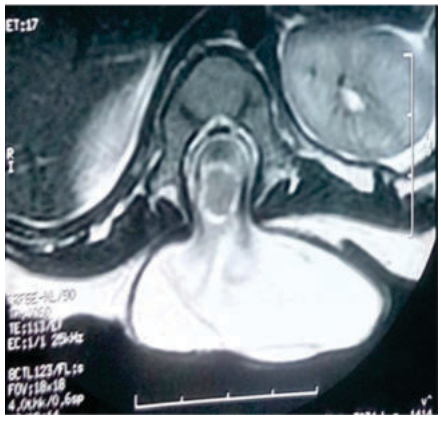
Spina Bifida is the most commoncongenital abnormality of the central nervous system , myelomeningocele being the most common anatomical type of spina bifida which comprises of approximately 85% of the lesions. Myelomeningocele is an entity which is common in the lumbosacral region due to congenital neurulation defect. The prevalence of myelomeningocele is approximately 0.8 to 1 per 1000 live births worldwide. The presentation is commonly a midline anomaly in the lumbosacral region(2). However, the anatomical location may vary as cases of cervical, cervicothoracic, thoracic, thoracolumbar, lumbar and sacral have also been documented. The sac usually contains nerve fibres or neural tissues, however sometimes it may contain intradural lipoma or dermoids.

Case Report

1 year 6 month old child presents in OPD with complaints of swelling over the back since birth. The baby was born with a small swelling over back which was initially the size of a peanut (1*1 cm) and gradually increased with age. The swelling increased on coughing and crying. There was no history of any discharge from the swelling or any ulcer over the swelling. The mother also complained of symptoms of urinary and bowel incontinence and weakness of both lower limbs. The baby is the first born male child and in antenatal period mother was a booked case with no significant antenatal history The baby was delivered at 9 months of gestation ageby normal vaginal delivery. The baby cried at birth with no history of ICU admissions. The postnatal phase was uneventful The motor milestone of standing with support and standing without support was delayed.

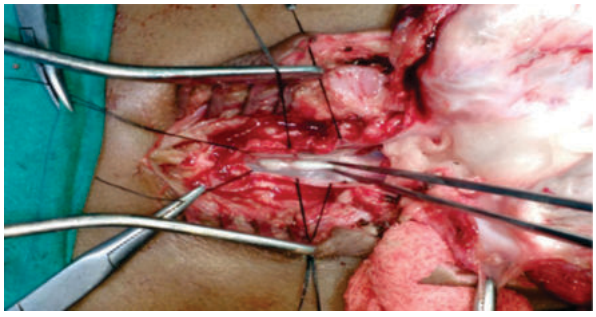
On systemic examination the the Nervous system revealed exaggerated deep and superficial reflexes. Other parameters were within normal limits. The local examinatiom of a back revealed a round tense cystic swelling of 8cm*9cm*6cm over the lower dorso-lumabar spine, of cystic consistency with fluctuation present at all axises, non-compressable, there is a gap in ythe spinous process of underlying vertebra. In the investigations the MRI revealed spinal dysraphism of D12 vertebra with meningomyelocoele and multi loculated syringomyelia.



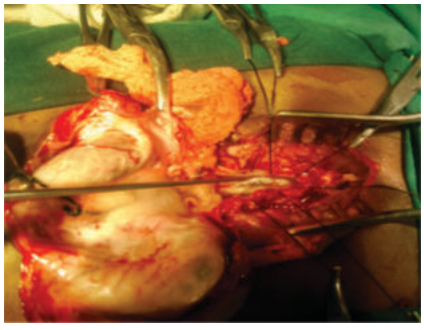


During the operative procedure of dethering and repair of meningomyelocoele sac, an epidermoid cyst was incidental found which was then excised. The procedure was done under GA in prone

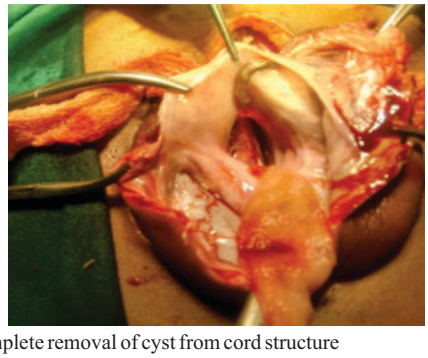
position. The histopathology showed cystic lesion lined by squamous epithelium containing keratin, which was consistent with the findings of an epidermoid cyst. On the three month follow-up of the patient he was found to be doing well from the neurological point of view.



Before Arachnoid dissection



After Arachnoid dissection



After complete removal of cyst from cord structure

DISCUSSION

The concurrence of a congenintal neoplasm within the spinal canal associated with myelomeningocele seems to be a very rare event as very few such cases have been reported worldwide. In 1894, Muscatello reported a female infant who had a thoracic meningocele and extradural epidermoid cyst at L4-L5 level, and a thoracic dermoid that was found at autopsy(3). In 1983 Martinez-Lage JF et al reported a case of a 4 month old boy with a lumbosacral meningomyelocoele that had an epidermoid cyst(4).

In 1966 Kirsch and Hodges reported epidermoid tumour with meningomyelocoele following surgical repair(5). In an article in 1993, Chadduck and Roloson reported a case of a newborn girl with myelomeningocele and an associated dermoid cyst in the filum terminale. The dermoid was found incidentally during myelomeningocele repair(6). The cases reported in 1994, 1972 and 2007 were teratomas associated with meningomyelocoele(7,8,9).

The origin of dermoid and epidermoid cyst is fundamentally the same. When invagination of skin elements into the neural canal takes place during embryonic life, a dermoid tumor will be formed and in case this occurs at a later stage an epidermoid tumor will develop. In our case we can rule out the traumatic origin as there was no history of spinal tap.

The association may be attributed to a common embryological origin for the occurrence of both the myelomeningocele and the dermoid as they can both be traced to the posterior neuropore closure at the 25 somite stage, at about the fourth week of embryonic life as stated by Martinez-Lage JF et al(4).

CONCLUSION

As per the literature the association of an extramedullary epidermoid in a case of myelomeningocoele is an extremely rare and uncommon occurence and possibly this is the only case to be reported in India to the best of our knowledge.

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