Case Report

Anatomy Section

A Rare Presentation of Fatal Sacral Meningocele: A Case Report

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ABSTRACT

A case of sacral meningocele presented as a diverticulum within the confines of a cystic enlargement found in a full term male foetus. The sacral meningocele does not cause any mortality, but in the present case the foetus is still born. This is the reason for taking up the case. An autopsy of the foetus revealed an extensive syringomyelia in the histological sections of the spinal cord at cervical, thoracic and lumbar segments.

Key Words: Sacral meningocele, Syringomyelia

INTRODUCTION

Meningocele implies a hernial protrusion of meninges through a defect in a skull or vertebral column Irving.s et al (1969) [1]. Intrasacral meningocele is an abnormal dilatation of meninges lying within confines of sacral spinal canal. Often the dilatation of sacral canal in all dimensions is so much that a thin bony wall remains Heppner & Diemeth (1960) [2].The cavity of meningocele communicates with subarachnoid space. Sacral nerves are found to be incorporated in the cavity.

CASE REPORT

A 30-year-old primi delivered a full term male foetus in the hospital. The dead foetus was brought to the department after the consent of the parents and protocol for preservation of the foetus was completed with 10% formalin. The foetus was born to the prime with no consangious marital history. A foetogram was taken before the foetus was subjected to fetal autopsy.

OBSERVATIONS

On fetal autopsy, an external examination of foetus revealed, CR length 29.6 cm. No morphological abnormalities were found except a cystic swelling of 15cm diameter occupying entire sacral region [Table/Fig-1].

An internal examination was conducted to find out any associated systemic anomalies in the abdomen and thorax. Nothing special was found out. Organs of all systems developed normally. The cranial cavity was opened for any anomalies of cerebral hemispheres, brain stem and cerebellum, nothing abnormal could be made out. After doing laminectomy from cervical to sacral region the spinal cord along with spinal meninges had been removed with the pedunculated cystic swelling of the sacral region. The cystic swelling was cut open for the presence of any embryological remnants of a teratoma. However nothing could be found. The cystic swelling was subjected to histological examination along with segments of spinal cord from cervical, thoracic and lumbar regions.

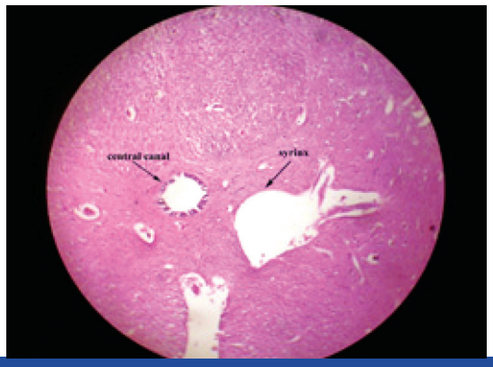
RESULTS

Foetogram revealed no abnormalities in vertebral column. The sections of spinal cord from cervical, thoracic and lumbar region are stained with haematoxylin , eosin and crystal violet. On histological examination, well developed multipolar cells in the grey matter of anterior horns have been observed, however around the central canal multiple syrinxes have been observed with dysgenesis of the parenchyma in the grey matter dorsal to the central canal. The spaces are seen as multiple cavities in serial sections of cervical [Table/Fig-2a], thoracic [Table/Fig-2b] and lumbar segments [Table/Fig-2c] that resulted in extensive syringomyelia of the spinal cord.

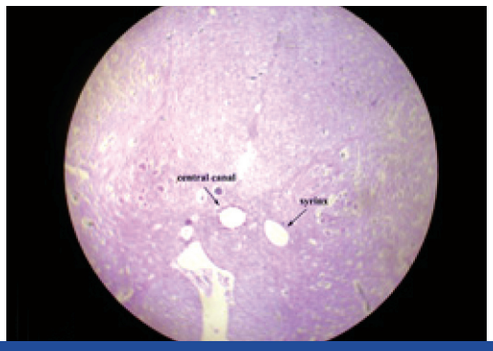
The cystic swelling of sacral region was composed of fibrous tissue covering with stratified squamous epithelium with mesodermal cells in clusters. Some of the longitudinal sections of the nerve fibres have also been observed which indicate that it is not a intrasacral meningocele [Table/Fig-3].

[Table/Fig-1]: Showing cystic swelling at sacral region

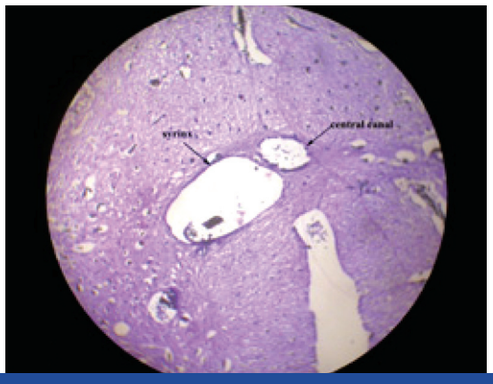
Spinal cord sections showing multiple syrinxes



[Table/Fig-2a]: Cervical region, H&E, 40 × 10



[Table/Fig-2b]: Thoracic region, crystal violet, 40 × 10



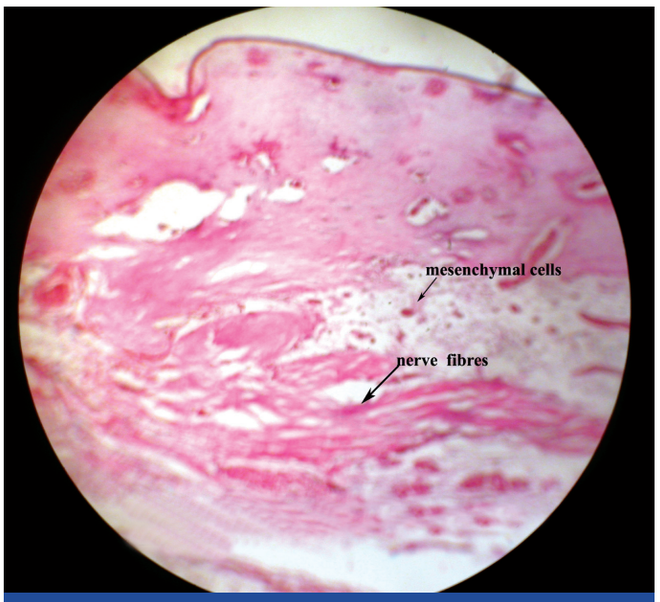
[Table/Fig-2c]: Lumbar region, crystal violet, 40 × 10

DISCUSSION AND CONCLUSION

Vasath. C, Talwakar, Darab K. Dastur 3 grouped meningoceles and meningo-myelocele into 9 subcategories according to anatomical lesion in the following order.

1A: Simple meningocele
1B: Meningocele with aberrant neural defect
1C: Meningocele with external fistula
1D: Meningocele with haemangioma
1E: Meningocele with tethered roots
1F: Meningocele with tethered cords
2A: Meningocele with ectopic spinal cord
2B: Meningocele with ectopic spinal cord and tethering
2C: Meningocele with total ectopia of spinal cord

The present case of cystic swelling in the sacral region come into the group 1E of Vasath C., Talwakar, Darab K. Dastur [3]. As the swelling was cystic with presence of recognisable nerve root and closely adherent fibres forming a part of the stalk. These roots formed a fibrovascular matrix of the stalk.



[Table/Fig-3]: Showing microscopic structure of meningocele, H&E, 10 × 10

Histopathological examination of supraradicular part of meningocele revealed only fibrous tissue and no smooth muscle and striated muscle collection. The above authors further describe inclusion of several varieties of congenital and pathological lesions involving spinal cord roots, meninges in thoracic, lumbar and sacral region.

Even though no evident radiological deformity had been found the present authors described a wide defect in a single spine that to lower lumbar and sacral regions with tethering of roots only which was observed in the present case also. Tethering of cord or roots to the undersurface of meningocele was probably responsible for neurological deficit in subgroup1E.

Bowton, Martin & Reckham (1966) [4] suggested the fibrofatty mass of sacral meningocele to be that of a haemartoma. Myelomeningocele also have been reportediy associated with other central nervous system anomalies like Arnold Chiari syndrome, hydrocephalus and syringomyelia (Punam singh et al. 2008) [5]. The authors have also described cerebral and cerebellar agenesis and dysgenesis of corpus callosum.

In the present case it is the syringomyelia that had been made out prominently and as it involved the entire length of cord region, the foetus could not survive because of extensive lesion. This may be the result of expression of unidentified development of gene or family of genes (Punam singh et al 2008) [5]. The authors also implicated that mutation for 5,10,5,10 methelenetetrahydrofolate reductase, C 67,77 allelic variant. The authors also mentioned that final confirmation is still awaited and warranted for further investigations.

To conclude the present case of fatal sacral meningocele is a very rare event. Antenatal diagnosis can be done with ultrasonography and measurement of amniotic fluid, alphafoetoprotein. It is to be measured out by pediatric neurosurgeon with the help of geneticist whether any intervention can be done antenatally in order to give the individual if at all the foetus survives an independent living status.

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