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CERVICOTHORACIC ARACHNOID CYST IN A PATIENT WITH NEUROFIBROMATOSIS: CASE REPORT
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B.B. SHEHU and I. HASSAN

SUMMARY

Intradural cervicothoracic arachnoid cysts are not common. They may be congenital, or secondary to trauma, surgery, haemorrhage, or inflammation. This is a report of a 39-year-old man who presented with cutaneous neurofibromatosis and cervicothoracic arachnoid cyst causing gradual quadriparesis. Magnetic resonance imaging showed an intradural extramedullary anterior cystic lesion at C_5 – T_2 level. Laminectomy and mersupialisation of the cyst was performed. Histology confirmed the diagnosis of arachnoid cyst. The patient recovered without neurological deficit.

INTRODUCTION

The occurrence of cervicothoracic arachnoid cysts is not common, (1-5) and arachnoid cyst occurring in association with neurofibromatosis is rarely reported. This is a report of a cervicothoracic arachnoid cyst in a patient with neurofibromatosis.

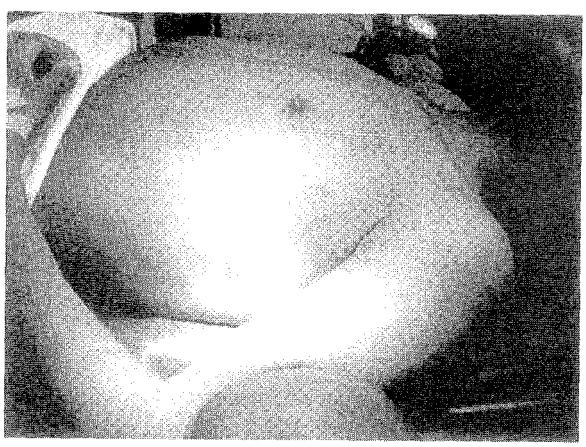
CASE REPORT

A 39-year old man presented with a one-year history of progressive quadriparesis with pain and hyperesthesia in both upper limbs. He could walk only with support. There was no urinary or faecal incontinence, and no visual or hearing impairment. There was history of trauma or spinal anaesthesia. There was no history suggestive of tuberculosis or meningitis in the past. No family history suggestive of neurofibromatosis or other neurological illness was obtained.

Physical examination showed widespread café au lait spots on the trunk (Figure 1); there were axillary freckles but no skin nodules. Examination of the eye and ears were normal. Glasgow coma score was 15 and all cranial nerves were intact. Muscle power was 4^+ in both upper limb muscles and 4^- in both lower limb muscles. There was hypertonia and increased deep tendon reflexes in both upper and lower limbs. Joint position sense was impaired in all limbs and there was hyperesthesia in both upper limbs. There was no spinal deformity or tenderness.

Figure 1

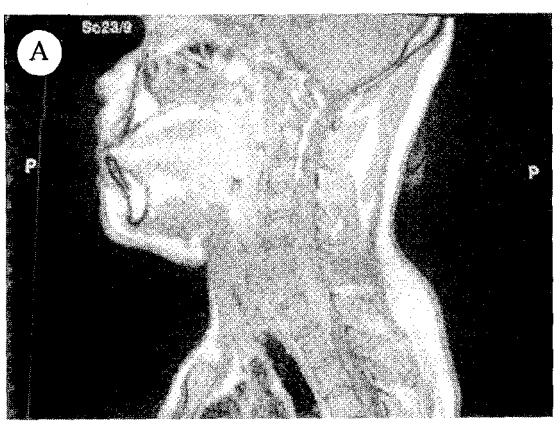
Café au lait spots on the patient's trunk

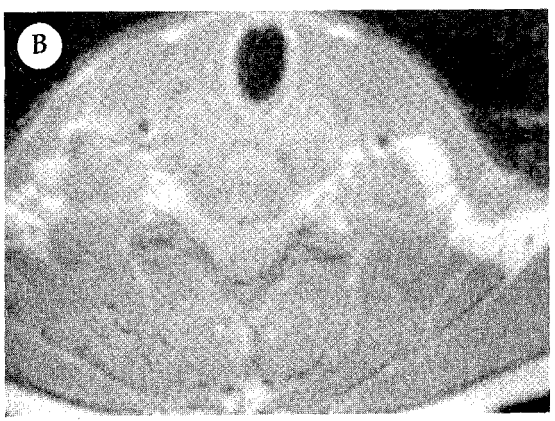


Complete blood count, erythrocyte sedimentation rate, Mantoux test, serum electrolytes and urea, and blood sugar were within normal limits. Cervical radiograph showed straightening of the cervical lordosis but there was no increase in the interpeduncular distance. Magnetic resonance imaging showed an anteriorly located intradural, extramedullary cystic lesion compressing the cord posteriorly, extending from C_5 - T_2 (Figure 2).

Figure 2

(A) Saggital cervicothoracic (T1 weighted) MRI showing a vertically located hyperintense intradural extramedullary lesion, extending from C_5 to T_2. (B) Axial (T1 weighted) MRI showing the cyst slightly hyper intense than CSF extending more to the right and compressing the cord.





At laminectomy (C_5 – T_2) a thickened, whitish arachnoid membrane filled with clear fluid was found; the spinal cord was displaced and flattened posteriorly. The cyst was marsupialised. Histology confirmed the cyst wall to be arachnoid membrane. The patient recovered full neurological functions and was discharged from hospital after three weeks. He has remained well at two years of follow up.

DISCUSSION

Arachnoid cysts are intra-arachnoid collections of cerebrospinal fluid (CSF). They may be congenital, acquired or idiopathic. They arise from accumulation of CSF within a split or duplicated arachnoid membrane. The CSF accumulates by secretion from arachnoid cells lining the cyst or trapped as a result of ball–valve mechanism in the cyst (2). Pathological distribution of arachnoid trabeculae leading to a diverticulum may also be an aetiological factor (6). The cyst wall consists of fibrous connective tissue slightly denser than normal arachnoid tissue (6). Secondary arachnoid cysts may be caused by inflammatory or infective diseases like arachnoiditis or meningitis, subarachnoid haemorrhage, trauma or spinal surgery. None of these aetiological factors was found in this patient. However, the café au lait spots and axillary freckles suggest neurofibromatosis (Figure 1).

Spinal arachnoid cysts account for 1 - 3% of all spinal tumours and occur most frequently in the thoracic spine (65%), then lumbosacral (13%), thoracolumbar (12%), sacral (6.6%), and cervical spine (3.3%) (6). Spinal intradural cysts are not common and rarely cause compression (7).

MRI is known to be the most effective means of making a diagnosis of arachnoid cysts; (5,8) it's non invasive and can be used for follow up. Cyst marsupialisation was effective in this patient and achieved full return of neurological functions. It's important to intervene early before permanent neurological deficits develops.

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