

Intramedullary pearly tumour

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Pearly tumours are benign and may occur at any age, but they are more common in young adults. The diagnosis of an intramedullary pearly tumour is rarely made before operation.

These tumours grow relatively slowly and the signs and symptoms as well as their rate of progression are related to the site of the tumour.

Pearly tumours are of congenital origin and their wall is composed of an outer layer of connective tissue and an inner layer of stratified epithelium (1). The centre of the tumour is filled with a cheesy mass of desquamated epithelium, sometimes mixed with cholestral crystals.

This content gives a shining pearly appearance to the tumour(2).

Treatment consists of surgical removal of the entire tumour with its capsule, but when it is in and intramedullary

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in position , only enucleation of the tumour and partial removal of its wall is possible(3).

An attempted total removal under such circumstances would probably result in a severe and permanent neurological catastrophe.

Recurrence tends to be slow in developing .

This report concerns two cases of intramedullary epidermoids, rarely described in neurological literature.

Case I:

The patient , a - 14 year old boy was first admitted on the 5th of Feb, 1969 giving a six year history of back pain . Two years after the start of this symptom he had developed numbness of the right calf, this being followed by slowly progressing weakness of the right foot and leg .

One year later similar symptoms had appeared in the left leg followed subsequently by sphincter disturbances .

On admission he was unable to walk and had pain in both legs. Examination disclosed nearly complete spastic paralysis of the legs and abdominal muscles. There was also disturbance of sensation in all modalities below the level of T6.

Reflexes were exaggerated in the lower limbs and Babinski sign was positive bilaterally.

The spinal X - ray was normal, and lumbar myelogram revealed total obstruction at the level of T6 . The protein content of the C.S.F. was 305 mg. per 100 ml. Exploratory laminectomy was performed.

It extended from the third to the seventh thoracic vertebrae.

The dure was seen to be under considerable tension without pulsation. After the cord had been split for a length of 3cm a pearly tumour appeared and was removed com -

pletely.

Post-operatively, the patient developed total paraplegia. It took the patient 14 months to recover with the help of physiotherapy.

The patient was seen again on Nov. 10, 1973 on account of numbness in his legs. The myelogram again revealed a blockage at the level of T6.

Following the myelogram, surgical treatment was suggested to the patient but he refused, as he was afraid of post-operative paraplegia. Symptomatic treatment did not help him, and he gradually deteriorated.

Subsequently, the patient consented to operative treatment which took place on April 13, 1974. At operation the dura was opened at the prior location. The tumour was covered with a thin layer of membrane. Excision of this membrane allowed removal of the tumour easily without damaging the cord.

Prior to this operation, the patient needed assistance for walking. Fortunately no deterioration was noted in the postoperative period. After a month, he could walk independently, but with some difficulty.

He has been in the outpatient clinic at regular intervals and there is no sign of recurrence. His gait improved further and he returned to his former employment (tailoring).

Case 2:

An 18 year old male whose symptoms appeared 4½ years prior to operation, was admitted to hospital on May 7, 1971. The symptoms were similar to the above case.

In this case the X-ray of the spine showed an enlargement of the spinal canal at the level of T9 to T10 lumbar myelogram revealed a complete block at this level.

The operation was performed on May 14, 1971 and as in the previous case, an-intramedullary pearly tumour was removed.

It took the patient 18 months to get rid of the sphincter disorders and to be able to walk with the help of crutches. Since last year, he has only been using a walking stick.

Discussion:

The intramedullary pearly tumour is a rare entity. Reviewing the literature, it appears to be responsible for 1-2 per cent of intraspinal tumours (4). In 301 intraspinal tumours at the Mayo-Clinic(5), there were four intramedullary pearly tumours(1.4%).

In our unit among 132 intraspinal tumours, we have had only these two cases of intramedullary pearly tumours(1.5%).

The prognosis is good, but sequellae may persist even years after the operation(6).

Summary:

The presentation treatment and postoperative progress of two patients with pearly intraspinal epidermoids is described. In one of the patients the tumour recurred after the first removal, necessitating a second operation.

Despite serious disturbances of gait both patients improved following surgery.

The literature in respect of these tumours is reviewed.

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