Case Report

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CRANIO-CERVICAL IDIOPATHIC HYPERTROPHIC PACHYMENINGITIS: A CASE REPORT AND REVIEW OF LITERATURE

Cranio-Cervical Idiopathic Hypertrophic Pachymeningitis: A Case Report and Review of Literature

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Abstract

Background Data: Cranio-cervical pachymeningitis is a rare and challenging disorder. Management of this condition is still under research.

Purpose: To follow a case of cranio-cervical pachymeningitis and to decide the optimal management method and factors affecting outcome and prognosis of this condition.

Study Design: A case report following surgical result with review of literatures.

Patients and Methods: A 56 years old male presented by a progressive cervical myelopathy. MRI cervical spine showed cranio-cervical hypertrophic pachymeningitis with compression on cervical cord. Surgical decompression was done with continuous postoperative clinical and radiological follow-up.

Results: Significant clinical improvement was observed after surgical excision. Recurrence happened later with cessation of corticosteroids. After follow up of the patient and review of literatures; factors affecting the prognosis were determined as follow: degree of surgical decompression, continuous use of corticosteroids till full radiological improvement, and development of cranial lesions.

Conclusion: Cranio-cervical pachymeningitis is a challenging disorder, management of it should be combined adequate surgical decompression, use of corticosteroids and postoperative rehabilitation program. (2016ESJ116)

Key words: Pachymeningitis, cervical spine, Inflammation, dura matter.

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Introduction

Hypertrophic pachymeningitis is a condition in which localized inflammation and thickenings of the dural matter happen. It can be divided into primary (with no definite cause) or secondary to other infectious or inflammatory disorders.[8] Secondary type is more common and happens as a consequence of one of the following disorders; inflammatory (Wegner disease, polyarterities nodusa , rheumatoid arthritis),[7] infective (neuro-syphlis, CNS tuberculosis, CNS cryptocossois, bacterial meningitis), neurosarcoidois, hemodialysis, and mucopolysacridois.[4] Primary (idiopathic) hypertrophic pachymeningitis on the other hand is a rare disorder with no definite cause.

Hypertrophic spinal pachymeningitis was first described by Charcot and Joffroy in 1869.[2] Its etiology still unclear, characterized by spinal cord pressure manifestations; progressive weakness, sensory loss, and sphincters un-control. It can also affect the cranio-spinal region with compression on the cerebellum, brain stem, and cervical cord with typically several levels of affection.

Management of this type of compression is usually surgical, the outcome, prognosis and factors affecting are still very under discussion because of rarity of the condition and paucity of literatures describing its proper management.

Patients and Methods

A 56 years old diabetic Sudanese male presented with a history of rapidly progressive quadri-myelopathy, he is presented in our clinic immobilized on a wheelchair. By clinical examination; motor power assessed by MRC grading scale,[11] Rt. side; foot dorsi-flexion and planter-flexion grade 0, knee and hip movements were also grade 0, Lt side on the other hand showed some better motor power; foot dorsi-flexion, grade 2, Lt. knee grade flexion and extension grade 2, hip grade 1. Upper limb also was more affected in left side rather than right side; left hand showed weakness grade 3, elbow flexion grade 2, Rt. Side hand and elbow were grade 1. Reflexes; exaggerated reflexes of both upper and lower limbs were grade 3.

MRI cervical spine showed longitudinal dural lesion extending form the posterior fossa in reteo-cerbellar region end extends caudally till the level of the third dorsal spine. The lesion was hyopintesne in T1-weighted images and also in T2-weighted images. The lesion was invading both anterior and posterior dura. It looked like an extradural lesion with an epidural collection or a tumor so it was confusing as an extradural lesion. However, with contrast enhancement the lesion looks more dural thickening configuration. It was taking segmental pattern, and the cervical cord was compressed in those segments. (Figure 1 A, B)

The patient had no history of previous tuberculosis, sarcoidosis or other chronic disease. Laboratory investigations were done; HIV, syphilis antibodies, tuberculosis, Torch, TB-PCR, tumor markers, complete blood count test, thyroid function, liver, and renal function. Her CRP, ESR, RF, ANA, antineutrophil cytoplasmatic antibodies (p-ANCA and c-ANCA), and all were negative. Surgical decision was taken.

Intra-venous high-dose methyl-prednisolone was taken as intravenous infusion on a rate of 5.4/kg/day for 2 days. This was started one day before surgery and continued through the operation and during the early postoperative period.

Surgical Procedure:

Under general anesthesia with fiber-optic intubation, posterior cervico- dorsal laminectomy from C3 up to D3 was done, with partial laminectomy of C2, no extra-dural lesions were found, dura was felt hard. Longitudinal incision was done along the whole length of the laminectomy. It was hardly thickened. Excision of the thickened parts was done as possible but not in all areas of the dura. Duoplasty with dural patch and fibrin glue was done. All layers are then closed tightly with mild-suction drain for 2 days.

Results

The excised dura was sent for bacteriological and histo-pathological studies but failed to identify a specific cause for this diffuse hypertrophy of the cranial and cervical dura. Histo-pathological analysis showed non-specific chronic inflammation with scattered collections of mononuclear inflammatory infiltrate comprising lymphocytes and plasma cell infiltration, fibrous tissue hyperplasia, hyaline degeneration. No granulomas or atypical cells seen, these were going was the typical appearance of idiopathic hypertrophic pachymeningitis described in literatures (Figure 2).[14] Cerebrospinal fluid (CSF) analysis was done, results were as follow; WBCs 85/cmm (lymphocytes), proteins 38 mg/dl, sugar 88 mg/dl and Gram's, Ziehl Neelson and India Ink stains were negative; no malignant cell was seen.

Postoperative Clinical Outcome:

The patient had a continuous physiotherapy program with a short course of corticosteroids and antibiotics. Patient showed significant improvement of motor power. He started moving all four limbs within one week and ambulated within a month. At that stage his motor power was improved significantly; in upper limbs; unfortunately, 3 months later, patient started to deteriorate again; clinical examination showed rapidly progressive deterioration till reach to the same clinical condition as preoperative.

Radiological Follow-up:

Follow-up MRI was not done immediately after surgery because of the clinical improvement. But, after recurrence of symptoms, new MRI with contrast was done and revealed recurrence of the compression patches at the same previous levels and also at a higher level on the occipital region compressing the brain stem and medullo-cervical region. CSF leak happened later because of wrong aggressive manual exercise for the neck (Figure 1 C).

Other surgical decompression was decided but patient was refusing and also he was showing gradual improvement after re-admission and starting IV corticosteroids, so surgery was postponed and cancelled at the end.

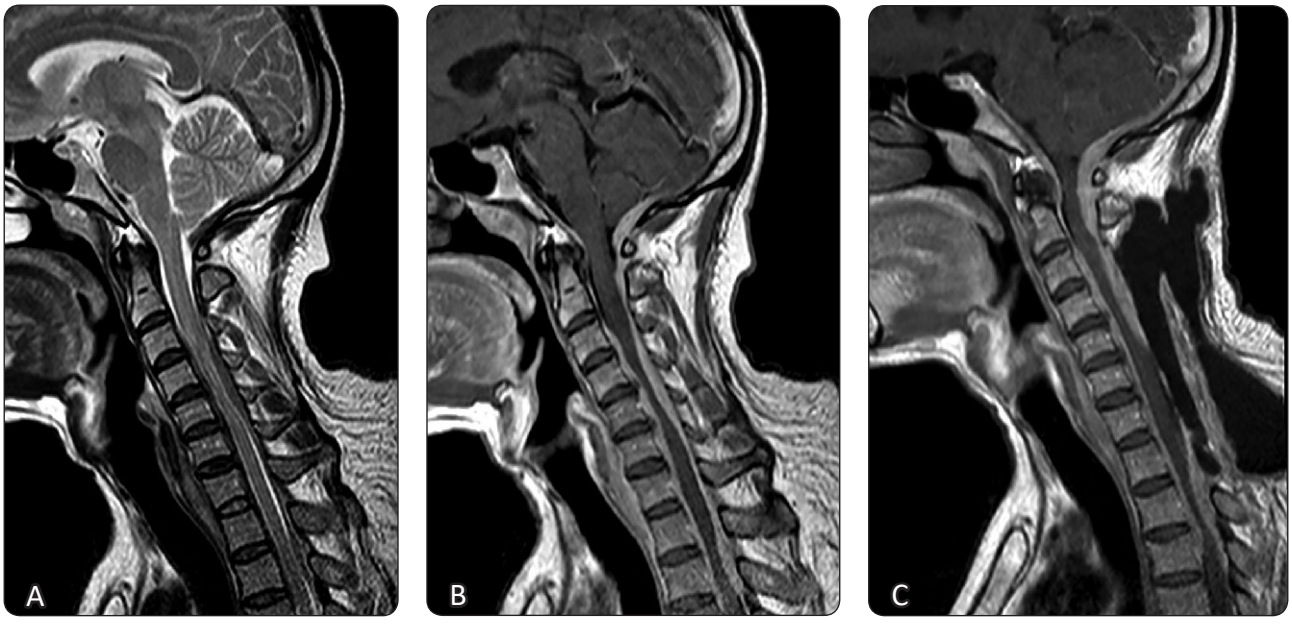

Figure 1. A) Sagittal view T2 WI of the patient showing patches of pachymeningitis involving the cervical and cranio-cervical regions. B) T1 WI with contrast shows more obvious views of the anterior and posterior patches compressing the cervical cord. C) Postoperative T1 WI with contrast showing recurrence of the cervical lesions with more increase in the size of the cranio-cervical lesion with more compression on the medulla oblongata, CSF leak appears. Areas of decompression also appear but insufficient.

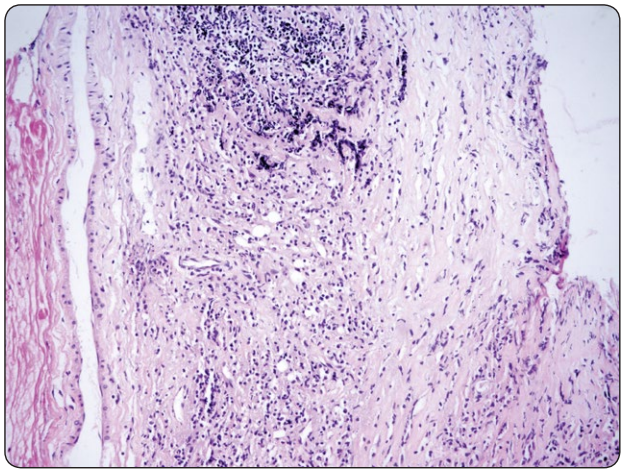

Figure 2. Pathology slide of the excised dura shows inflammatory cells with lymphocytes and plasma cells.

Discussion

Cranio-cervical primary pachymeningitis is a rare condition presented by neural compression manifestations. Spontaneous regression was described by many authors.[9] Management of this condition is surgical in most of the cases due to compression of the cervical cord. The usual technique is by longitudinal opening of the dura and excision of the thickened dura with release of the compressed spinal cord.[5] In certain circumstances, decompression is inadequate and the thickened dura circumferentially surrounds the spinal cord, so only posterior parts, and to some extent lateral parts can be excised but the remaining parts can shows progressive increase in thickness and more compression on cord.

Factors are essentially studied to be predictors of bad prognosis during the surgical treatment of cervical pachymeningitis: Inadequate decompression, No long-term corticosteroids.[15] Aggressive course of progression: the course of this disorder is unpredictable, so strict postoperative follow-up is required at least for the first year after surgery. This should be done clinically and radiologically, and development of cranial lesions

In a review of a similar cases of cranio-cervical pachymeningitis; Bottella et al,[1] discussed similar surgical approach and observed a temporary improvement only. In his study, prognostic factors were not clear. We tried to mention surgical factors for prognosis; the most important looks to be the degree of decompression and the use of corticosteroids.

Khadilkaret et al,[9] in a case study about surgical treatment of cervical idiopathic pachymeningitis, he diagnosed the case by exclusion of other secondary disorders that can be a causative factor. He surgically treated the case by posterior decompression but his case showed relapses and remissions for 5 years duration on intermittent courses of corticosteroids.

Takahashi H et al,[13] operated a case of ventral cervical pachymeningitis by anterior cervical decompression and fusion, with marked neurological improvement after surgery and corticosteroids. The affected site showed no remission on MRI with continuous steroid therapy.

Most authors agreed that the use of corticosteroids is the cornerstone of treatment of hypertrophic pachymeningitis and some had marvelous results and total remission of the lesion with use of immunosuppressant factors such as methotrexate. Other physicians on the other hand agree that corticosteroids only have a partial effect at the beginning of treatment but have no effect later-on, and relapses will happens on just stop of medications.[10] In our case study, we observed relapse of the case after stoppage of corticosteroids, that is why we agree that early discontinuation of corticosteroids can lead to relapses. We can recommend continuing use of corticosteroids till full resolution of the lesions in MRI images with a more safety period for assurance.

Surgical treatment was tried by many authors[3,6] relapses can occur and can be managed again by corticosteroids only. Surgical resection as possible, high dose of

corticosteroids, continues strict follow upon this condition clinically and radiologically, and administration of corticosteroid on recurrence.

The outcome of surgical excision seems to be dependent on the invasiveness of the cervical dura from anterior and posterior directions. The extent of surgical excision with a wide laminectomy and removal of the whole posterior dura with duroplasty and the application of fibrin glue could be important factors of decreased recurrence incidence. Again, this should be combined with continuous postoperative corticosteroids till full disappearance of the lesion in MRI.

Conclusion

Cranio-cervical pachymeningitis is a challenging disorder, management of it should be combined adequate surgical decompression, use of corticosteroids and postoperative rehabilitation program.

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الملخص العربي

التهاب بطانة السحايا التضخمي في منطقة اتصال الرقبة بالجمجمة. تقرير عن حالة مع مراجعة الأبحاث

البيانات الخلفية: التهاب بطانة السحايا التضخمي هو اضطراب نادر وصعب. ولا تزال طريقة علاج هذا المرض قيد البحث.

الغرض: لمتابعة حالة من التهاب بطانة السحايا التضخمي وتحديد أسلوب العلاج المثلى والعوامل التي تؤثر على النتيجة والتشخيص لهذا المرض.

تصميم الدراسة: متابعة نتائج عملية جراحية مع مقارنة النتائج بالأبحاث المنشورة سابقا عن نفس الموضوع.

المرضى والطرق: متابعة حالة رجل يبلغ من العمر ٥٦ عاما قدمه يعاني من اعتلال النخاع العنقي التدريجي. أظهر التصوير للعمود الفقري العنقي بالرنين المغناطيسي وجود التهاب بطانة السحايا التضخمي مع ضغط على الحبل العنقي. تم رفع الضغط جراحيا مع المتابعة السريرية والإشعاعية المستمرة بعد العملية الجراحية.

النتائج: لوحظ تحسن سريري كبير بعد الاستئصال الجراحي. حدث تكرار في وقت لاحق مع وقف الستيروئيدات القشرية. بعد متابعة المريض ومراجعة الأدب. تم تحديد العوامل التي تؤثر على التشخيص على النحو التالي: درجة من الضغط الجراحي، والاستخدام المستمر من الستيرويدات القشرية حتى تحسين الإشعاعي الكامل، وتطوير الآفات الجمجمة

الاستنتاج: لالتهاب بطانة عنق الرحم يمكن أن يكون إلزاميا في حالة ضغط الحبل الشوكي كبير. يعتبر استخدام الستيروئيدات القشرية حجر الزاوية لعلاج هذه الحالة. مستقبل الحالة يمكن تحديده من قبل عوامل معينة.