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Intramedullary spinal cord cavernoma

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Abstract :

The interamedullary spinal cord cavernomas are uncomman.A 48 years old female presented with progressive weakness and numbness of both lower limbs followed by gradual neurological deterioration with bladder symptoms. MRI revealed an intra medullary dorsal cord angiomatous lesion. A complete microsurgical removal of the cavernoma was carried out.

Histopathology revealed it to be cavernous haemangioma. The clinical, radiological features andtherapeutic strategy of Intramedullary Spinal Cord Cavernoma (ISCC) are discussed.

Keyword :

cavernoma, Intramedullarycavernoma, spinal cord cavernoma,ISCC

Introduction:

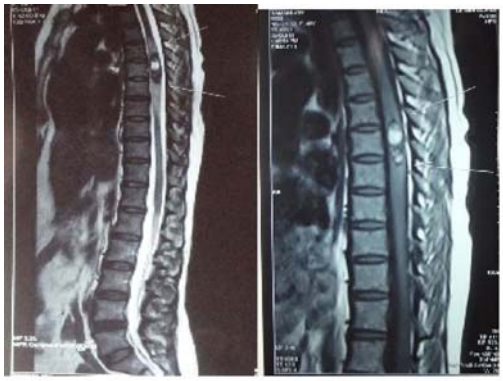
Spinal cavernomas are rare. Spinal cavernoma constitute 5% to 12% of spinal vascular abnormalities are well circum-mscribed lesions that consist of closely packed, sinusoidal spaces, without intervening spinal tissue. Hadlich reported the first definitive account of ISCC in 1903 and Schultze reported the first total resection of an ISCC in 1912. More ISCC have been detected through extensive use of MRI, based on his surgical results McCormick first advocated complete resection of ISCCs in 1988 [(2)].

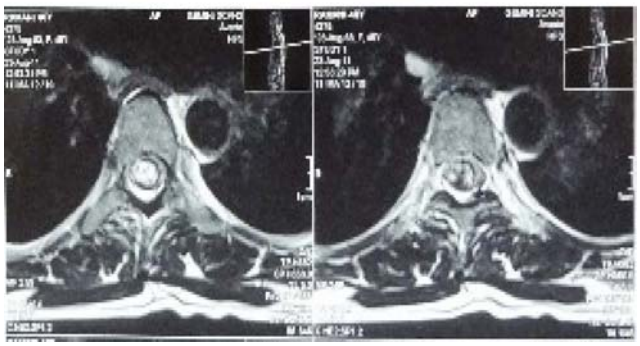
Case report:

A 48 years old female had difficulty in using both lower limbs in the form of tripping of toes and buckling of knees and benumbed sensation below the hip for the past one month. 20 days later she had dull aching back pain and progressive-worsening of weakness in both lower limbs, inablity to walk associated with stiffness and also had difficulty in squatting, getting up from squatting position.She had also incontinence of urine and constipation. Neurological examination showed

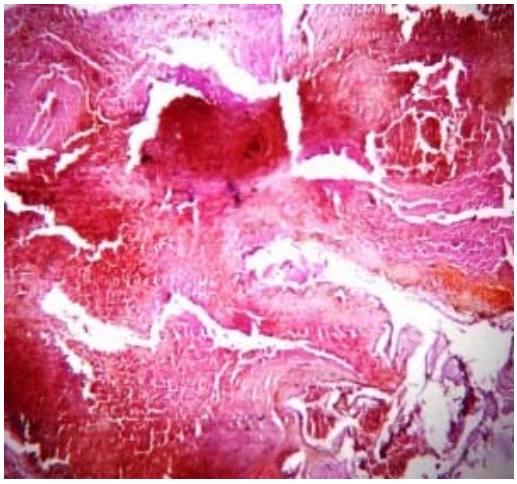
hypertonia in both lower limbs.The power in right lower limb was grade 3 and left lower limb was grade 2, pyramidal type of weakness.The deep tendon reflexes both side knee jerks were exaggerated with bilateral ankle clonus and abdominal reflexes absent in all quadrants and plantar bilateral extensor. Graded sensory-loss of all modalities touch, pain, temperature, vibration and joint position sense below D8. No spinal tenderness or deformities.

MRI of dorsal spine revealed an intradullary lesion at D6, which was heterogeneously low to iso intense with central hyper intensity onT1W1 and hyper intense onT2W1 with peripheral hypo intensehaemosiderine rim with late sub acute hemorrhage(Figure1,2,3).





Dorsal laminectomy was planned, D5, D6& D7 laminectomies were done. After the durotomy, under the operating microscope a blusih black discoloration was seen on left side of spinal surface at the level of D6. The myelotomy was done in left paramedian side of the bluish black looking discoloration of the spinal cord. The total micro surgical excision of the mass was performed by dissecting in haemosiderin



stained gliotic plane. After excision of the mass the dura was closed completely. Themass was dark blue mulberry shaped consisting of multiple cyst with old clotted blood.

Histopathological examination of the mass revealed the presence of sinusoidal vascular spaces lined by endothelial cells, characteristic of cavernous haemangioma(Figure.4). After surgery, the patient was relieved of pre operative back pain. From the third post operative day the patient showed improvement in sensation of pain, touch and temperature. After tenth post operative day, the power in both lower limbs became 3 plus. Bladder and bowel symptoms didn't improve.

Literature Review:

Intramedullary cavernomas are relatively rare, but recent MR imaging studies have demonstrated that they are more common than originally thought. Cavernoma induced myelopathy was misdiagnosed as idiopathic myelopathy because these lesions were difficult to identify on computerized tomography and myelographyMore recent estimates that cavernomas constitute 5% to 12%

of vascular malformation. The literature review revealed that the mean age at symptomatic presentation and diagnosis is usually in 3rd or 4th decade of life, occur more frequently in females with approximate ratio of 2:1 and the thoracic cord is most frequently involved , followed by the cervical cord[3].

Radiological appearance:

Cavernomas have a characteristic appearance on MR images. They have well circumscribed, mixed high and low signal intensity on T1-weighted images. Hemosiderin deposits cause a low signal intensity surrounding ring on T2-weighted images. Contrast enchancement is slight or nonexistent[4].

Pathological characteristics Cavernomas consist of closely packed sinusoidal spaces and do not contain intervening spinal tissue. They have no significant venous drainage and no large vascular supply. On gross inspection, cavernomas have a mulberry appearance and haemosiderine stained. Microscopically they are characterized by thin walled sinusoidal vascular channels [1].

Clinical presentation:

Presented with progressive paraparesis, sensory loss and frequently associated pain. It may be indistinguishable from chronic progressive radiculomyelopathy.This varied manifestation is attributed to its variable size, which may vary from few millimeters to several centimeters. Acusymptoms are probably caused by new haemorrhages within or around the lesion. Slowly progressive course of the cavernoma may be due to local pressure effects on adjoining spinal cord and or repeated episodes of bleedings. Since the haemorrhage in cavernous haemangioma is venous at low pressure, the presentation is not as dramatic as with arterial bleed. Incidental asymptomatic lesions have also beenreported. The neurological deterioration may have sometimes variable degree of recovery although most patients exhibit a gradual clinical decline. On histo-pathological correlation, four major clinical p atterns have been defined: (a)acute episodes of stepwise deterioration with small but repeated haemorrhages or with thrombosis of malformed vessels, (b)slow progression due to progressive enlargement of the cavernoma eventually with thickening of the sinusoid vessels and gradual thrombosis, (c)acute onset with rapid deterioration due to intraparenchymal haemorrhages, (d)acute onset with gradual decline attributable to altered microcirculation due to intraparenchymal haemorrhages[5].

Surgical Management:

The annual bleeding rate of ISCCs is estimated to be between 1.4% and 4.5%.cavernomas have a significant rebleeding rate associated with further neurological decline[2]. These vascular malformations can also lead to a progressive myelopathy that can be arrested by surgery.Cavernomas are usually amenable to surgery. A gliotic plane usually develops between the lesion and the spinal cord, allowing for relatively easy removal of the lesion. The lesions usually come to the dorsal surface, perform a simple laminectomy. A myelotomy is made directly over the discolored area where the cavernoma comes to the dorsal cord surface.he cavernous malformationis then removed microsurgicaly in an inside out fashion until the gliotic white matter plane. To achievetotal removal one should examine the operative bed more carefully to identify any residual lesion that might give rise to further recurrence. According to some reports transient neurological worsening immediately after surgery is seen in almost half of patient and is mostly followed by a gradual recovery. If the patient suffers an acute decline in neurological function due to acute largehaemorrage, surgery should be performed

4-6 weeks later, when a gliotic plane developed between the lesion and spinal tissue[2].

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