DUMB - BELL GANGLIONEUROMA PRESENTING WITH THORACOLUMBAR SCOLIOSIS (A CASE REPORT)

A. M. Alpaslan *, E. Acaroğlu **, M. Kış **, O. Çataltepe ***

A case of an 11 year old girl who has three years history of deformity on her back with no neurological symptoms is presented. Initially diagnosing the disease as juvenile idiopalhic scoliosis, a two staged procedure consisting of an anterior instrumentation and fusion as a second stage, were planned. In spite of the fact that there was a block at L3 level at lumbar myelographic examination, her CT scans were interpreted as mechanical obstruction due to spinal deformity. A retroperitoneal mass, extending into the spinal canal through intervertebral foraminae in a dumb-bell fashion, could only be seen at the first stage operation. Microscopic examination of the tumor indicated a ganglioneuroma. Laminectomy of nine vertebral levels had to be performed so as to resect all epidural extensions of the mass.

At her three months follow-up, she is still wearing a Risser Cast with no trace of reoccurrence on CT scan.

Key Words: Thoracolumbar Scoliosis, Ganglioneuroma.

A wide variety of histologie types of paravcrtcbral tumors are known to have the ability to grow through intervertebral foramina in to the spinal canal. Of tumors originating from the neural crest, neuroblaslomas with such extensions causing deformities and neurologic problems have been reported (1,3.4). Such an extension of a ganglioneuroma, appears to be very rare.

Ganglioneuramos arc fully differentiated tumors of neural crest that contain no immature elements. They are quite rare compared to other benign neural tumors but they are known to outnumber neuroblastomas along the sympathetic axis 3 to 1. Majority of them are diagnosed in second and third decades of life, most favorable location being posterior mediastinum followed by retropcritoneum. Metastatic spread of gangl ioneuromas arc extremely rare and arc due to malignant transformation (2).

The first case of a dumb-bell ganglioneuroma was reported by Lorets in 1870 which was diagnosed in a postmortem study. In 1958 Shephard and Sutton reviewed the literature and found 12 such cases and included their own four cases. Of the 16 cases 11 had complete paraplegia, the others had neurologic deficits of varying extent but only one of the patient was known to have a spinal deformity (5).

Since then only a few such cases appeared in the literature known by us. In 1989 Bauer ct al. reported a case of an 16 year old girl who had been followed up for four years as progressive idiopalhic scoliosis and signs of paraparcsis (1).

We hereby report our case of an 11 year old girl with 3 years history of deformity and no neurologic involvement.

CASE HISTORY

The patient was first seen at our hospital in 1988 at the age of 10. She had two years history of deformity on her back. A right thoracolumbar scoliosis was diagnosed. On radiologic examination her Cobb angle from T 11 to L5 was 32° (Fig 1) She was called back 6 months later and it was seen that her deformity had increased to 40° (Fig 1) A year after initial diagnos is she complained of rapid progression of her deformity and her Cobb angle was found to be 72° (Fig. 1) A surgical approach, consisting of two stages, first anterior release, instrumentation and fusion, then posterior release, instrumentation and fusion, was planned.

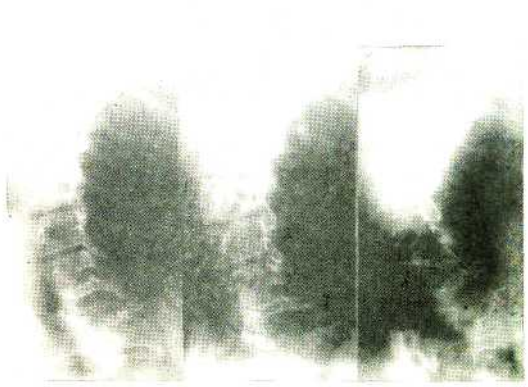

Fig 1.

* Professor, Hacettepe University Medical Faculty Dept. of Orthopaedics and Traumaiology.
** Research Assistant, Hacettepc University Medical Faculty Dept. of Orthopaedics and Traumatology.
*** Research Assistant, Hacettepe University Medical Faculty Dept. of Neurosurgery.

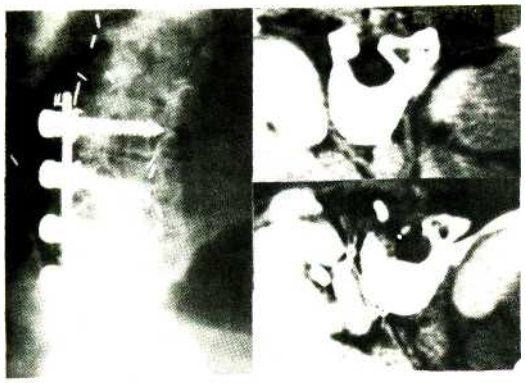

Fig 2.

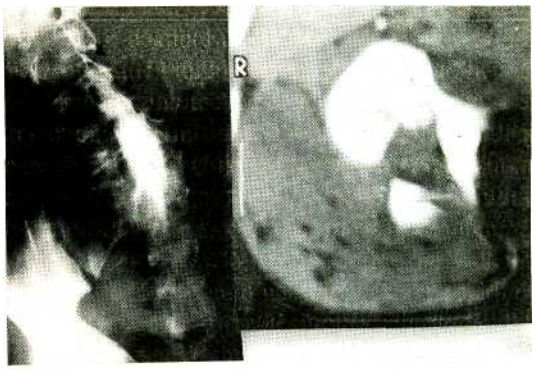

Fig 3.

Physical examination at the time revealed a right throcolumbar scoliosis with a hump at right lower thoracic region. Her pelvis was tilted to right and her right lower extremity was 2 cm longer than left measured from umblicus. She had no loss of motor power or sensation, her anal sphincter tonus was normal.

Radiologic examination showed a 72° thoracolumbar scoliosis from T11 to L5 which improved only 8° on traction or lateral bending.

An iohexol myclography was performed via a puncture in the L4-5 interval and a block to the passage of contrast medium was seen at L3 level. (Fig.2) Only a small amount of dilute contrast material could be traced up to LI level. A computed tomographic examination with contrast myclography was performed but, because the images obtained were all in bone density the cause of the obstruction was thought to be mechanical due to the deformity of the spine. (Fig.2)

A yellow-gray, well encapsulated, rubber like mass, 8x5x5 cm in size was encountered at the first stage of her operation in right rctropcritoncal region and traced up to Ll-2-3-4 intervcrtcbral foraminac. The paraspinal portion of the mass was excised and Wcbb-Morley instrumentation performed. When the pin was being placed into the body of T12 a fracture of both of the pedicles occured with a very minor force and the T12 corpus fractured. On examination, it was seen that, the A.P. diameter of the corpus was only 1.5 cm. Neural canal was inspected from the defect and it was seen that the epidural space was full with the same ncoplastic tissue pushing the cord to the left and anterior.

Histologic examination of the specimen revealed the tumor to be a ganglioncuroma.

At the second stage, it was found that the intracanalicular portion of the mass extended from the level of L5 up to T9 so lamincctomy of nine vertebral levels had to be performed so as to resect all the remnants of the tumor. Postoperative roentgenograms following a two-stage procedure reveal that the scoliosis has been reduced from 72 to 26 degrees. (Fig. 3)

Three months follow-up showed no change in her neurological status and CT scans revealed no residual or recurrent mass. (Fig. 3)

DISCUSSION

Our case illustrates a very unusual cause of thoracolumbar scoliosis. It could only be diagnosed during surgical intervention. A retrospective" review of the case shows that there were indeed some hints of an extra ordinary reason for the deformity. A rapid progressive lumbar curve have suggested a deformity due to spinal disraphism, compared to typical curve patterns of idiopalhic scoliosis. That being the fact, the CT scans could have been analysed in soft tissue densities, looking for a spinal tumor or congenital abnormality. But,, since the patient was free of neurological symptoms, and radiologic examination showed no trace of congenital deformity, we did not look for another etiologic factor. As we have stated above, ganglioneuromas arc benign tumors, their treatment being simple resection. The life expectancy of our patient appears to be normal. A review of the literature suggests that the prognosis of paraspinal ganglioncuromas with dumbbell extensions are poor due to the neurologic deficites they cause. But it should be noted that most of the cases reported have been diagnosed and treated in the first half of the century when evaluation of such tumors and deformities were extremely difficult. Since the case presented was caught before any neurological symptoms occurred and since the tumor was totaly ex-

cised, it is reasonable to expect a favourable prognostic outcome.

It is necessary to stabilize the spine prior to development of rigid postlaminectomy deformities because of performing laminectomy at the nine levels. However, since at the first stage operation anterior instrumentation and fusion were performed at T12-L4 levels, no posterior instrumentation was considered at the second stage. For the time being, we are planning to wait for the anterior graft consolidation and see if any instability demanding surgical correction will take place in the patient's future follow-up.

REFERENCES

1. Bauer B.L., Bauer H., Griss P., Lutcke A., Maroske D., Mennel H.D., Unsicker K.: Dumb-bell ganglioneuroma of the spine misinterpreted as progressive idiopathic scoliosis. Arch. Orthop. Trauma Surg. 108: 189-94,1989.
2. Enzinger F.M., Weiss S.W. (Eds): Soft tissue tumors: 828-31, The C.V. Mosby Company; 1988.
3. King D., Goodman J., Hawk T., Boles T., Sayers M.P.: Dumb-bell neuroblastomas in children. Arch. Surg. 110: 888-91, 1975.
4. Klimmer M., Hecker W.Ch., Zur Klinik Von Neuroblastomen und Ganglioneuromen mit intraspinaller betciligung. Z.Kinderchir. 43: 343-6, 1988.
5. Shcphard R.H., Sutton D., Dumb-bell ganglioneuromata of the spine with a report of four cases. Brit. J.Surg. 45(192): 305-17,1958.