HEMANGIOBLASTOMA CAUSING CERVICAL NEURAL FORAMINAL WIDENING

I.G. BILGEN (1), N. YÜNTEN (1), E. DEMIRTAS (2), S. ÇAGLI (3), M. ZILELI (3)

(1) Ege University Faculty of Medicine, Departments of Radiology Bornova — 35100 Izmir, Turkey.
(2) Pathology.
(3) Neurosurgery.

SUMMARY

We present the MRI findings in a case of a 24-year-old woman with spinal hemangioblastoma, causing neural foraminal widening by producing a dumbbell mass in the lower cervical region. Hemangioblastomas can very rarely present as an intradural extramedullary lesions and this case is another exceptional pathology which should be considered among the differential diagnosis of enlarged intervertebral foramen due to neoplastic processes.

Key words: spine, neoplasms, hemangioblastoma, magnetic resonance imaging.

CASE REPORT

A 24-year-old woman was admitted to the hospital with a 1-year history of progressive numbness and weakness in the left hand, and left sided hemiparesis. Physical examination revealed left dominant spastic quadriparesia and left sided deficit of sensation at the C5-6 level.

Conventional radiography showed widening of the left C5-C6 neural foramen (figure 1). Cervical MRI revealed a dumbbell mass showing extraradial extension into the left scaleneus muscle. The left neural foramen at C5-C6 level was prominently enlarged (figures 2a, 2b). The mass had heterogeneous signal on T1 (figure 2a) and T2 (figure 2b) weighted images and showed prominent foci of high velocity signal loss. Diffuse and marked enhancement was noted on postcontrast T1-weighted images (figures 3a, 3b). The C5 vertebral body showed left posterolateral erosion due to the compression effect of the mass. The neighbouring vertebral artery, mildly displaced ventrally, was patent. On coronal images, the extradural component of the mass descended down to the C6 level (figure 3b). On T2 weighted axial images, the spinal cord was compressed and displaced to the right. Thoracic MRI showed an intradural round mass lesion of 1 cm diameter at T4 level, which showed marked enhancement on postcontrast images.

The masses, which were resected partially at the cervical and totally at the thoracic regions, were found to be highly vascular at surgery. Histopathological examination of both tumors revealed a hemangioblastoma. They were composed of two components, large variably vacuolated stromal cells and a rich capillary network (figure 4). The stromal cells had round to so-
mewhat irregular nuclei and eosinophilic or variably lipid-rich cytoplasm. The cells were uniformly distributed within network of capillaries, reticulin strain showed honey combed pattern (figure 5). Mitoses were absent. Stromal cells were vimentin positive, epithelial membrane antigen and glial fibrillary acidic protein negative. KI-67 PI was 0.1 %.

Von Hippel-Lindau syndrome was excluded by a negative family history, and a normal cranial CT, abdominal ultrasound (US) and eye examinations of the patient.

DISCUSSION

Among various neoplastic, vascular and developmental causes of spinal neural foraminal widening, the most common pathology is the dumbbell schwannoma and neurofibroma [1, 3]. Rare causes include congenital absence of pedicle [1, 5], lateral meningocele [1, 6], vertebral artery aneurysm or tortuosity [1, 4], tuberculous abscess [6], and rare tumors such as chordoma, dermoid, meningioma, chondroma, chondrosarcoma, neuroblastoma, lipoma and malignant fibrous histiocytoma [1-3, 6-8].

Hemangioblastomas are rare, accounting for 1 % to 5 % of all spinal cord tumors [9, 10]. They may be encountered at any age, but are usually seen in young adults (the average age is 30 years) [11, 12]. Seventy-five percent of hemangioblastomas are intramedullary, and 10 % to 15 % are both intramedullary and extramedullary masses. Extramedullary hemangioblastomas are often attached to the dorsal spinal cord pia and are very rare [12, 13]. Approximately one third of patients with spinal cord hemangioblastomas have von Hippel Lindau (VHL) syndrome. Retinal or cerebellar involvement typically precedes spinal cord symptoms in these patients [9]. In our patient, both abdominal US and eye examinations were normal. Family history and
screening for visceral manifestations of VHL disease were negative. So our case was regarded as a « sporadic case ».

Macroscopically, a typical spinal cord hemangioblastoma has a highly vascular nodule with a large cystic component that diffusely enlarges the cord. Prominent leptomeningeal vessels are usually present. Microscopically, spinal hemangioblastomas are characterized by densely vascular tissue that consists of thin walled, closely packed blood vessels interspersed with large pale stromal cells [14].

On MRI, hemangioblastomas show diffuse cord expansion with high signal intensity on T2W images, and prominent foci of high velocity signal loss. The tumor nodule in the spinal cord enhances strongly following contrast administration [9, 12, 15].

Our experience with the present patient demonstrates that a rare form of a hemangioblastoma, originating at an intradural and extradural compartment, can pass through a neural foramina, thus causing neural foraminal widening and producing a dumbbell lesion. In our opinion, hypervascularity of the...
tumor on MRI examination is a main, prominent feature that can contribute to the diagnosis in such cases. In the differential diagnosis of dumbbell lesions and neural foraminal widening, hemangioblastomas should also be considered especially when prominent foci of high velocity signal loss is present.

REFERENCES


