Intramedullary capillary hemangioma associated with hydrocephalus in an infant

Case report

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This 3-month-old child presented with an enlarging head circumference arising from communicating hydrocephalus with large subarachnoid spaces in the posterior fossa. Neuroimaging performed to clarify the origin and pathogenesis of the hydrocephalus revealed a vascular lesion within the dorsal spinal cord. Insertion of a cerebrospinal fluid shunt and total removal of the spinal tumor were performed successfully. Histological examination of the medullar lesion demonstrated a capillary hemangioma. Proposed mechanisms for increased intracranial pressure and spinal cord lesions are presented. A spinal hemangioma in this age range associated with hydrocephalus has not been reported previously, but spinal lesions must be considered in the presence of hydrocephalus with no clear origin.

KEY WORDS • intramedullary hemangioma • hydrocephalus • spinal cord • blood vessel neoplasm • pediatric neurosurgery

S PINAL cord hemangiomas are known as rare benign lesions, are described as occurring in the adult population in the fifth to sixth decade of life, and have no sex predilection. The most frequent location is in the lower thoracic segment, conus medullaris, and cauda equina,11 but more recently the involvement of the upper thoracic cord has been reported.2–4 Spinal hemangiomas in pediatric patients have never been described, although they are probably hamartomatous in nature.5 The association of spinal lesions with hydrocephalus is well known, but the pairing almost exclusively concerns parenchymal tumors as astrocytomas and ependymomas, or, more and more rarely, vascular tumors. To our knowledge, only one case of a spinal hemangioblastoma associated with hydrocephalus has been reported.6

The infant we describe in this paper is therefore the first known pediatric patient affected by a medullar capillary hemangioma and hydrocephalus; in addition the association of spinal cord hemangioma and hydrocephalus has not been reported until now.

Case Report

History and Examination. This 3-month-old boy was admitted to the Pediatric Neurosurgery Unit of the University of Pisa because of an abnormal growth in head circumference. His mother's pregnancy was uneventful, and the baby was born at full term with a head circumference of 36 cm at birth, 43 cm at 1 month of age, and 48 cm at 2 months. At presentation, physical examination revealed a head circumference of 49.5 cm and a wide, full, tense anterior fontanel with diastatic sutures. Mild irritability and lethargy were also apparent. No other neurological signs were present.

Neuroimaging. A CT scan of the skull revealed a marked dilation of the ventricular system, with large subarachnoid spaces in the posterior fossa. An MR image of the brain confirmed these anatomical abnormalities (Fig. 1). Because the cause of the condition was not evident, a spinal MR imaging study was performed. A high thoracic (T4–7) intramedullary lesion was discovered that was isointense on T1-weighted images with vascular high-flow components, and enhanced markedly after Gd administration (Fig. 2).

Surgical Procedure. Because of the clinical signs of marked intracranial hypertension, a ventriculoperitoneal shunt was placed initially, which controlled the hydrocephalic symptomatology well. A CSF sample obtained from the ventricular cavity had normal protein, glucose, and cell-count values. At the end of this first surgical procedure, before the child was awakened, a spinal puncture allowed the examination of the CSF of the spinal segment: again, glu-
cose and cell count values were normal, whereas protein levels were 60 mg/dl (the high limit of the normal range).

After 1 week, a second surgical procedure was performed: the spinal lesion was approached through a dorsal laminectomy. After opening of the dura mater and the arachnoid membrane, a subpial medullary tumor was detected on the dorsal surface of the cord; dilated vessels were noted into and on the lesion. The tumor was dissected progressively from its lower pole in the plane between the lesion and the parenchymal cord, with meticulous coagulation at low power and cutting of all feeding vessels with microscissors (Fig. 3).

**Histological Analysis.** Histological examination revealed a lesion composed of many capillary-sized vessels lined by normal endothelial cells; no cavernous elements were present. The stroma were constituted by a fine network of reticulin fibers; a few foamy cells were detected, but no neuron-specific enolase or $S100$ immunoreactivity was observed. A histological diagnosis of capillary hemangioma was made (Fig. 4).

**Postoperative Course.** The postoperative period was uneventful; the child was discharged 7 days after surgery with no neurological deficits. After 1 year, he is neurologically intact, with no focal deficits and with normal psychomotor development. Cerebral and spinal MR imaging verify the regression of the ventricular dilation and the absence of lesions in the spinal cord.

**Discussion**

Capillary hemangiomas are not rare lesions, but they are more frequently observed in the skin or other soft tissues. In contrast, spinal intradural hemangiomas are reported only sporadically and occur exclusively in adults. Spinal cord hemangiomas are exceptional. Only seven cases have been described to our knowledge, and they were localized mostly in the lower segment of the thoracic spinal cord. Ours is the first observation of medullary hemangioma in an infant, even if this lesion is considered hamartomatous in nature and was presumably present until birth. The lesion was clinically silent, but the association with a communicating hydrocephalus allowed the diagnosis.

Spinal neoplasms and communicating hydrocephalus in the same patient is a rare but well-known event; children younger than 3 years of age, as was our child, may be affected more frequently. Generally, spinal lesions associated with hydrocephalus are neoplastic in nature, in particular intramedullary, low-grade gliomas of the spinal cord. Different mechanisms have been proposed to justify this association. The authors of one series proposed that the involvement of the cervicomedullary junction and obex may block the CSF circulation, but their hypothesis is valid only for spinal tumors that extend in this region. Other authors suggest the intracranial spread of tumor cells through the subarachnoid pathways, but this “neoplastic arachnoiditis” cannot explain the hydrocephalus observed with benign tumors, as in the most frequently described cases, or in the presence of vascular spinal lesions. A high level of CSF protein content, even if transitory, may justify the associated hydrocephalus; this increase of proteins may be secondary to the lesion itself because of the following: 1) a chronic aspecific inflammatory reaction to the lesion; 2)
an alteration of the blood–brain barrier at the level of the tume
toral vessels;15 or 3) chronic microscopic hemorrhage into
the subarachnoid space from the lesion, especially if vas-
cular in nature. A protein migration from the vessels of the
lesion to the CSF space may be caused also by a local im-
pairment of blood circulation because of venous hyperten-
sion in the case of vascular lesions. Once proteins have
induced an increase in CSF outflow resistance because of
the induced basal arachnoiditis, fibrous adhesions, and con-
sequent obliteration of CSF spaces, hydrocephalus may
persist even if the protein level returns to normal values. In
the case reported here, the hemangioma was discovered be-
fore the onset of any spinal signs or symptoms. In explain-
ing the presence of communicating hydrocephalus with-
out a sure origin, we cannot exclude the possibility that the as-
sociation is fortuitous, as suggested by some authors.3

Because of the marked intracranial hypertension ob-
served in our patient, a ventriculoperitoneal shunt was in-
serted at first to control the communicating hydrocephalus;
the removal of the spinal tumor only has been reported to
be sufficient to obtain the resolution of hydrocephalus in
some cases,16 but this is not a common observation.6

The diagnosis of a spinal tumor can be delayed if the
clinician’s attention is focused on the earlier and often
marked signs and symptoms of intracranial hypertension;
in our case, the need to pinpoint the origin of the hydro-
cephalus correctly allowed an early diagnosis of the spinal
lesion before it became symptomatic. Surgical treatment
of the spinal lesion was performed in the best clinical con-
ditions, resulting in no neurological deficits.

Once the diagnosis of a spinal dorsal lesion was ob-
tained, our next diagnostic hypothesis was hemangioblas-
toma, because of the characteristics discerned on MR imag-
ing. The lesion was isointense on T1-weighted images, had
high-flow vascular components, and promptly enhanced af-
after Gd administration. It is worthwhile noting that a correct
preoperative diagnosis was not made in the unique series
reported in the literature either.5 At surgery the lesion ap-
peared as a reddish tumor of moderately hard consistency;
only microscopic examination allowed the diagnosis of
capillary hemangioma because of the paucity of the foamy
stromal cells that are plentiful in hemangioblastomas,2,12
and the absence of neuron-specific enolase and S100 im-
munoreactivity.19 According to Roncaroli, et al.,18 we think
that the differentiation of capillary hemangioma and he-
mangioblastoma can be based on histological examination

FIG. 2. Spinal MR images. A: Sagittal T1-weighted image depicting the intramedullar isointense lesion. B: The le-
sion is markedly enhanced after Gd administration. C: A coronal image confirming the lesion; in all images flow-void
signals are present in the tumor, confirming the presence of high-flow vessels. D: Axial contrast-enhanced image dem-
onstrating the dorsolateral location of the mass into the spinal cord.
only. The distinction between these two vascular lesions is mandatory because a hemangioblastoma can be the sentinel of von Hippel–Lindau syndrome, with a different prognosis and the need for strict follow up. Hemangiopericytomas often show cystic components, but because of the positive neuron–specific enolase immunoreactivity of these tumors, only histological examination can provide a sure diagnosis. In all cases, neuroradiological screening is advisable because of the possibility of multiple hemangiomas, even if hemangiomatosis is observed only occasionally.

The surgical goal is the total removal of the hemangioma (recurrence has never been reported). During surgery, bleeding should be avoided whenever possible; this result may be achieved by not entering the lesion. Manipulation of the spinal cord should be minimized to avoid surgical injuries to nervous tissue and its supplying vessels. Dissec-

Fig. 3. Intraoperative views. A: The lesion is detected on the dorsal surface of the spinal cord; dilated vessels are present on it. B: Surgical removal of the tumor is completed.

Fig. 4. Photomicrograph. Capillary hemangioma highlighting a network of blood vessels lined by normal endothelium in a fibrous stroma. H & E, original magnification × 20.
tion in the plane between the tumor and the spinal cord with low-power coagulation is mandatory to obtain a good postoperative outcome.

Conclusions

Capillary hemangiomas are rare tumors of the spinal cord; however, they should be considered when neuroradiological examinations show vascular lesions in the spinal cord because surgical removal is possible without inducing new neurological deficits.

References


Manuscript received August 28, 2004. Accepted in final form April 28, 2005. Address reprint requests to: Aldo Iannelli, M.D, Institute of Neurosurgery, University of Pisa, Via Roma 67, 56126 Pisa, Italy. email: a.iannelli@ao-pisa.toscana.it.