

Cervical Ganglioneuroma and Obstructive Hydrocephalus Following Surgery - A Rare Association

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Abstract

Ganglioneuromas are rare tumors, known as neuroblastic or neurogenic tumors, which most often start in autonomic nerve cells, which may be found in any part of the body.

We present a case of 53 years old, woman, with mild arterial hypertension, who has right sided hemiparesis, more on the leg, with gradual onset. Cerebral MRI was normal.

After a few months, she developed motor deficit on the opposite leg, with the picture of tri paresis and impairment of walking. MRI of the cervical spine was showing an extra medullary intradural mass C5-C6-C7 and she finally underwent extended cervical laminectomy from C-5 to C-7 level, and total ablation of the tumor. Despite the first supposition of a neurinoma, histopathology examination confirmed a case of ganglioneuroma tumor.

In a few weeks, she became confused and with impairment of gait. An obstructive hydrocephalus was confirmed on CT scan. The ventriculo-peritoneal shunt was performed and the recovery was complete.

Keywords: Ganglioneuroma; Dumbbell shaped; Cervical spine; Active hydrocephalus; Motor deficit; Intradural mass

Background

Ganglioneuromas are rare tumors, known as neuroblastic or neurogenic tumors that most often start in autonomic nerve cells.

This type of rare tumors tends to occur in the pediatric population and is often asymptomatic. They are occasionally diagnosed in adults due to their mass-effect on adjacent structures [1].

Usually, ganglioneuromas are considered the most mature form of neuroblastoma, originating from neural crest cells that normally migrate into the adrenal medulla and sympathetic ganglia.

The retro-peritoneum and posterior mediastinum are the two most common locations for these tumors, followed by the cervical region of spine [2].

Case Presentation

We present a case of 53 years old, woman, with mild arterial hypertension, who has right sided hemiparesis, more on the leg, with gradual onset. Cerebral MRI was normal. After a few months, she developed motor deficit on the opposite leg, with the picture of tri paresis and impairment of walking. MRI of the cervical spine was showing an extramedullary dumbbell shaped mass at the C5-7 level extending outside through neural foramen.

Neurologic examination reveal spastic gait Babinski sign bilaterally present, osteotendinous reflexes slightly elevated except left tricipital reflex, global hypoesthesia at C5 level, more on the left side, episodic

urinary incontinence. MRI examination of cervical spine shows us cervical intradural mass (58/28 mm) at C5-C6 level, T1 isointense, T2 hyperintense, with non-homogeneous gadolinium enhancement, with multiple cystic like zones, left paravertebral extension and vertebral scalloping C6-C7, well capsulated, which pushes the spinal cord laterally, being well delimited from it (Figures 1-4).



Figure 1: Before surgery - Sagittal T2.



Figure 2: Before surgery - Sagittal T1+contrast.



Figure 4: Before surgery – Sagittal STIR.



Figure 3: Before surgery - Sagittal T2.

Our patient accepted neurosurgical treatment. After laminectomy, the neurosurgeon described extradural and intradural dumbbell shaped tumour more than 80% of medullary channel and with an extrapleural extension to left pulmonary apex. The ablation of this tumor was complete; duration of surgical intervention was about 420 min.

The incision was cervico-thoracic, posterior medio-spinal occipital-thoracic, curved to the left reaching T2 level, with cervical laminectomy C5-C7 and osteosynthesis with spongyous grephon. After the dura was incised, the compression of the spinal cord was observed. This occupied approximately 20% of the rachidian canal. The tumor was extra-intradural, dumbbell appearance, with extension to C6 left foramen reaching the extrapleural left pulmonary apex.

Macroscopically the tumor was white-yellowish with hard consistency, with cystic areas with moderate bleeding.

Anatomo-pathological examination established the type of tumour being ganglioneuroma, with intense hemorrhagic areas.

The MRI evaluation of the cervical spine after surgery revealed posterior paravertebral and intrachannellar lichenoid mass (5.47/2.55/5.11 cm) with no gadolinium enhance, with caudal extension reaching subcutaneous level and postero-cranial reaching the spinal apophyse, without gadolinifile areas. The medulla was compressed and pushed to the right, antero-laterally (Figures 5-8).



Figure 5: After surgery - Sagittal T1.



Figure 7: After surgery 1 month - Sagittal T1 contrast.

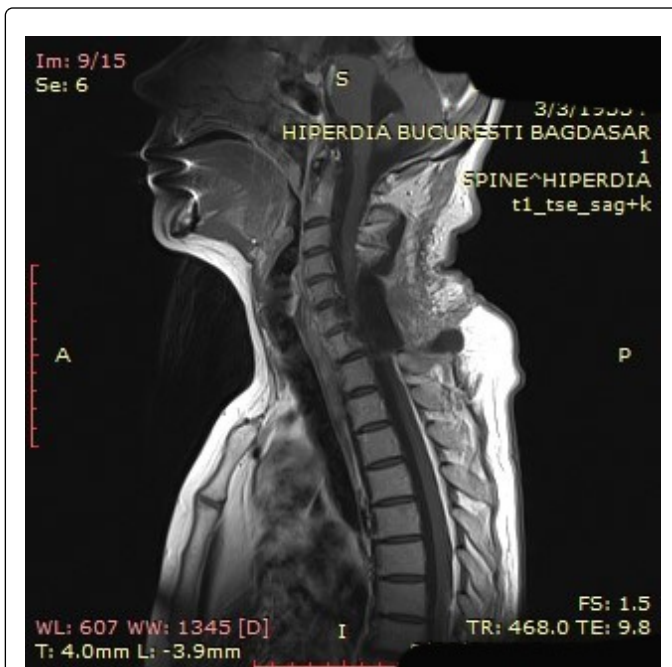


Figure 6: After surgery - Sagittal T1+contrast.



Figure 8: After surgery 1 month sagittal T2.



Figure 9: Computer tomography showed active hydrocephalus, with enlargement of all ventricles.

Neurological exam was normal at discharge from hospital, except slightly elevated osteotendinous reflexes.

After three months, the patient came to the hospital with vomiting, decreased mentation, confusion, astasia-abasia, impairment of gait with insidious evolution.

Computed tomography of the brain showed marked dilatation of ventricles suggesting active hydrocephalus and low density of the periventricular area due to transependymal CSF resorption (Figure 9).

CSF analysis was conducted before surgery, with normal macroscopic appearance. Microscopic examinations of CSF, including routine cell counts and biochemistry, were normal.

A surgical intervention was performed and ventriculo-peritoneal shunting procedure was performed, with a Delta Medtronic low pressure device implanted. The patient was discharged from the hospital fully conscious and with no neurologic deficits.

After one year, the patient has a normal neurological exam, cerebral CT scan normal, with patent shunt; MRI of the cervical spine has the same aspect (pseudo-meningocele like cervical mass).

Discussion

Spinal tumors occur with an incidence rate of 1.1 per 100,000 persons. Intramedullary spinal tumors comprise approximately 2-4% of all central nervous system neoplasms. The most common kinds of intramedullary tumors are ependymomas, astrocytomas and hemangioblastomas.

Nerve sheath tumors (NSTs) constitute approximately 25% of all tumors arising in the intradural extramedullary space [3].

The majority of the spinal tumors reported in the literature are of neural lineage including schwannoma. Intramedullary tumors are lesions that usually arise directly from the neural tissue of the spinal cord. The differential diagnosis of the most common presenting signs and symptoms include intradural extramedullary spinal tumors, epidural spinal tumors, myelopathy due to degenerative disease, cord infarct, vascular lesions such as spinal arteriovenous malformations and dural arterio-venous fistulae, the inflammatory processes such as multiple sclerosis, transverse myelitis and sarcoidosis [4].

Ganglioneuromas in adults are rarely reported, but final diagnoses were histopathological. MRI is the modality of choice for evaluating

the extension of spinal tumors. In general, neuroblastic or neurogenic tumors appear radiologically as well-circumscribed, smooth or lobulated masses that may contain calcifications. Dumbbell appearance of spinal tumors refers to a tumor which has both a component within the canal and a component in the paravertebral space linked by tumor traversing the neural exit foramen. The appearance of dumbbell tumors is related with different types of tumors [5], spinal nerve sheath tumors (schwannoma, neurofibroma or ganglioneuroma), spinal meningioma or neuroblastoma.

The benign (ganglioneuromas) and malignant (ganglioneuroblastomas) forms of these tumors are virtually identical radiologically. The only differentiating factor is the possibility of distant metastases with malignant ganglioneuroblastomas.

Complete surgical resection of ganglioneuromas is important, because it allows for good tissue sampling and a thorough pathology examination of the specimen to ensure a correct diagnosis of ganglioneuroma. In rare cases, these tumors recur; therefore, radiological examination is an important tool for the proper localization and characterization of primary and recurrent tumors.

We found a similar case with a ganglioneuroma located at cervical level of spine, C4-C5, with positive outcomes and no other complication [6]

Cervical pseudomeningocele after surgery is not such a common complication but in this case is related with obstructive hydrocephalus. It is more difficult to differentiate ganglioneuromas from schwannomas or neurofibromas. In general, schwannomas and neurofibromas are round and can cause bone erosion and destruction. Most ganglioneuromas are flat and elongated and they normally do not affect the bone. Neurofibromas do not have a capsule; the presence of one in a patient would point to a ganglioneuroma diagnosis [7].

Hydrocephalus was related to spinal tumors and surgery at this level, but usually after decompression for Chiari Malformation or cervicomedullary junction surgery. The mechanism of developing hydrocephalus is secondary to obstruction of the CSF outlets [8-10]

Conclusion

We presented a rare case of ganglioneuroma at adult age, with post-surgery pseudomeningocele and obstructive hydrocephalus. As a particularity for this case, we established the age of the patient and

location of the spinal tumor (C5-C7). The patient has no neurological deficits for the moment.

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