Hemothorax presenting as a primitive thoracic paraganglioma

Case illustration

STEFANO TELERA, M.D., MARIANTONIA CAROSI, M.D., VIRNA CERASOLI, M.D., FRANCESCO FACCIOLI, M.D., EMANUELE OCCIPINTI, M.D., ANTONELLO VIDIRI, M.D., AND ALFREDO POMPILI, M.D.

Divisione di Neurochirurgia, Divisione di Chirurgia Toracica, Servizio di Radiologia e Diagnostica per Immagini, e Servizio di Anatomia Patologica, Istituto Nazionale Tumori “Regina Elena,” Roma, Italia

KEY WORDS • primitive thoracic paraganglioma • hemothorax

Primitive paragangliomas of the thoracic spine are rare lesions that may arise from paraganglia cells of the spinal sympathetic chain. They contain catecholamines in cytoplasmic granules, but they are seldom active in neuroendocrine secretion.1,2,4

This 59-year-old woman with recurrent back pain presented with respiratory distress after suffering an acute episode of sharp pain involving the left hemothorax. Chest radiography revealed a significant left pleural effusion. Thoracic computerized tomography (CT) and magnetic resonance (MR) imaging demonstrated an inferior lobe atelectasis of the left lung and a large extra- and intracanalicular tumor at the level of T10–11, which eroded the intervertebral foramen (Fig. 1). The pleural effusion proved to be hemorrhagic during thoracentesis. Whole-body CT scanning showed no signs of abnormality.

The patient underwent radical surgery via a left thoracotomy along the 10th rib and a costotransversectomy. The pleura and lung were covered by a film of old blood. The lesion had a well-defined cleavage plane and a rich vascular supply from both the intercostal arteries and aorta. Considerable intraoperative bleeding occurred during resection; this made the procedure demanding and required that the patient be given multiple blood transfusions.

The histological diagnosis was paraganglioma (Fig. 2). Because the levels of catecholamines and their metabolites in plasma and urine were normal, the lesion was classified as a nonfunctioning paraganglioma. The postoperative course was uneventful. Four months after surgery, MR imaging confirmed complete removal of the tumor and disappearance of the pleural effusion. At the 14-month follow-up examination there was no sign of disease recurrence, and whole-body CT scanning documented no other lesions.

To the best of our knowledge, including our patient, only 14 cases of primitive thoracic paragangliomas have been reported. The tumor was extradural in 12 cases and intradural in two. In two patients local recurrence was demonstrated; cerebrospinal fluid (CSF) or distant metastases occurred in three patients.1,2,4 These features are different from those of the more common paragangliomas of the lumbar spine and cauda equina, which are mostly intradural and are never associated with metastases outside the central nervous system; CSF dissemination was described in 1% of these cases.1

Acute onset of symptoms due to hemothorax has not been previously reported in lesions involving the thoracic spine. Hemorrhage secondary to the hypervascularized tumor may explain the pleural effusion in our patient, because large extradural paragangliomas in other locations are known to become hemorrhagic and necrotic.3

The possibility of local recurrences or distant manifestation of other paragangliomas, even after a long period, emphasizes the value of prolonged postoperative surveillance.1,2,4

Fig. 1. A: Axial T2-weighted MR image revealing a well-demarcated T10–11 lesion in the paravertebral space with intraforaminal component and extension to the costovertebral junction; the lesion exhibited mildly hyperintense signal intensity. B and C: Axial and coronal spin echo T2-weighted images obtained after contrast medium administration, showing an intense enhancement in relation to high vascularity. The left lung is covered with blood degradation products.

Fig. 2. Photomicrograph of the thoracic paraganglioma specimen showing nests of closely packed polygonal cells with small round-to-oval nuclei and clear-to-faintly eosinophilic cytoplasm surrounded by a delicate fibrovascular stroma. Nuclear pleomorphism without conspicuous mitotic activity can be appreciated. Immunoreactivity was positive for synaptophysin, neuron-specific enolase, chromogranin, and S100 protein (not shown). H & E, original magnification ×20.

References


Manuscript received July 7, 2005; accepted in final form March 21, 2006.

Address reprint requests to: Stefano Telera, M.D., Divisione di Neurochirurgia, Istituto Nazionale Tumori “Regina Elena,” Via Elio Chianesi 53, 00144 Roma, Italia. email: telera@ifo.it.