CASE STUDY

Multiple Ganglioneuroma With Cervical Involvement

Ganglioneuromas múltiples con afectación cervical

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Clinical Case

A 62-year-old male, who was a regular smoker and drinker, presented at the ENT Department with a laterocervical tumour which had evolved over several months. There were no clinical symptoms involved, except local irritation in the area and a gradual increase in tumour size.

Examination revealed a 7×4 cm right laterocervical mass at levels II–III, which was elastic in consistency and nondisplaceable; there were no signs of inflammation.

Nothing of significance was observed on examination of the oral cavity. A nasofibrolaryngoscopy revealed right pharyngeal swelling, at the same level as the cervical tumour protrusion and we found no other pharyngolaryngeal lesions or alterations in vocal cord motility.

The cervicothoracic CT scan (Fig. 1A) showed a lesion with the same density in its soft parts, with hyperdense foci, and extensive peripheral calcification, measuring 4.4×3.3×5.1, located on the right postrolateral level, posterior to the ipsilateral internal carotid artery. There were no other notable cervical findings. However, in the last thoracic incisions, the presence of another tumour with identical characteristics was discovered, attached to the back wall of the gastric corpus, measuring 5×4×4.3. For this reason, we extended the study of the abdomen (Fig. 1B), where this finding was confirmed.

By means of an ultrasound guided FNA, amorphous material with calcification was obtained, and with no epithelial cellularity, but it did not provide sufficient information to confirm the disease. In the absence of diagnostic guidelines, we considered it imprudent to make an incisional biopsy, and we therefore requested a cervicofacial MRI and a CT guided percutaneous biopsy, together with an assessment by the Internal Medicine Department.

The imaging test did not provide any new data, and the material extracted did not contain sufficient cellularity to establish a histological classification. The broad study performed by the Internal Medicine Department, including serology and bacilloscopy, ruled out any infectious aetiology.

Figure 1 (A) Cervicothoracic CT scan showing heterogeneous lesion with large-scale peripheral calcification, posterior to the internal carotid artery. (B) Abdominal CT scan showing lesion next to the posterior gastric wall.

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Figure 2  (A) Ganglioneuroma resected with internal carotid artery scarring on its anterior side. (B) Fibrous stroma showing ganglionic type cells with clear nucleolus and granular cytoplasm with pigment on the inside.

With no suspected diagnosis, we considered the need for excision of one of the lesions. We chose to perform a cervicotomy, since the patient was abdominally asymptomatic and the considerable size of the cervical tumour caused progressively more severe discomfort.

A cervicotomy was performed under general anaesthesia, with the removal of a specimen of $8 \times 6 \times 5.5$ (Fig. 2A). It was brown in colour and of very irregular consistency, tightly attached to the posterior side of the carotid artery. The vagus nerve and the internal jugular vein were contained inside the mass, and were therefore removed. On incision, we noted multiple calcifications which were heterogeneous in appearance, interspersed with haematologic areas with other nodules. Extemporaneous histological analysis (Fig. 2B) established a diagnosis of ganglioneuroma (GN) of the sympathetic cervical chain which contained cysts and secondary calcification.

Catecholamine production by GN is less frequent than in neuroblastomas; however, we did rule out the raising of homovanillic acid and vanillylmandelic acid serum.

At present, 9 months after surgery, the patient is presenting with sequelae of Horner syndrome and paralysis of the right vocal chords. They are being followed-up by the General Surgery Department. For the time being, in the absence of symptoms no abdominal surgery is indicated.

Discussion

GN are benign tumours of the peripheral nervous system which comprise mature ganglion cells and Schwann cells, and they stem from the neuroectodermal crest. They usually present in patients aged between 10 and 30, with a slight predominance in females. They may be found anywhere in the sympathetic chain, but the usual location is the posterior mediastinum (41%) and the retroperitoneum (37%), with infrequent cervical presentation (1%–5%). Complications are regularly defined and they are asymptomatic until their size leads to manifestations of the disease due to the mass effect on nearby structures.

Although they are infrequently seen in this area, GN have to be included in the differential diagnosis of cervical masses. Due to low frequency and lack of specific signs and symptoms, it is often difficult to reach a diagnosis prior to a histological examination. CAT scans and MRI scans provide valuable information on size, location and association with neighbouring structures, such as the large cervical vessels, with the subsequent prevention of severe lesions in surgery. However, it is still difficult to discriminate GN from other cervical lesions. The use of FNA is frankly limited and the only method of diagnostic confirmation is surgical excision, also taking into account the possibility of clinically presenting malignant neoplasms and with similar radiological characteristics such as neuroblastomas or ganglioneuroblastomas. Possibly the only way of suspecting this diagnosis is a thorough knowledge of GN and keeping them in mind as a possibility.

In our case, complexity increased due to the particular age of the patient and the unusual existence of another abdominal tumour. Although cases of multiple GN have been described in the cervical area, multiple GN in 2 distinctive anatomical locations are extremely rare.

Studies have been made regarding the concurrence of neuroectodermal crest tumours in patients affected by type I neurofibromatosis, although their higher frequency than in the general population has only been confirmed in pheochromocytoma. In this case, neither the clinical symptoms nor examination were compatible with the existence of neurofibromatosis.

Treatment of GN consists of complete surgical resection, and the patient should be warned of the probable neurological sequelae. Due the possibility of local recurrences there is a need for periodic imaging testing during follow-up.

Conflict of Interest

The authors have no conflicts of interest to declare.

References