CASE REPORT

Intrathoracic schwannoma with Horner syndrome

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ptorpiano@gmail.com**SUMMARY**

Horner syndrome (HS) results from the interruption of the sympathetic pathway to the eye and face, and describes a collection of signs consisting of ipsilateral miosis, partial ptosis, anhidrosis and apparent enophthalmos. It is a clinical observation, and has a plethora of possible causes, ranging from the benign to the malignant. Involvement of the stellate ganglion on the sympathetic chain by malignant tumours of the lung is a well-recognised cause of HS. On the other hand, HS secondary to the excessive growth of a benign intrathoracic neoplasm is a very rare finding, with only a few cases described in the literature. Our patient was found to have such a diagnosis when he presented to medical attention with a 1-month history of cough that was associated with features of HS that he had ignored for the preceding 9 years.

BACKGROUND

This case illustrates the rare occurrence of Horner syndrome (HS) secondary to a benign intrathoracic schwannoma, and exemplifies the importance of thoroughly investigating any patient with HS.¹ It also highlights the possibility of mistaking stridor for wheezing, as what initially was managed as an acute asthmatic exacerbation, unresponsive to nebulisers, eventually resolved following surgical resection of an intrathoracic schwannoma, with no further exacerbations in the follow-up period.

CASE PRESENTATION

A 32-year-old Caucasian man presented to the emergency department with a 1-month history of cough productive of white sputum and associated with progressive shortness of breath on exertion, which had persisted despite courses of co-amoxiclav and moxifloxacin. He was a known asthmatic, taking inhaled beclomethasone and salbutamol. Despite this, he smoked about 15 cigarettes every day. He had not noticed any haemoptysis or weight loss.

On examination, his vital parameters (temperature; oxygen saturation; blood pressure; respiratory rate; pulse rate) were all within the normal range for a man of his age. Auscultation of the chest revealed reduced air entry in the right upper zone, with occasional expiratory wheezing. He was also noted to have right-sided HS (figure 1). On questioning, he explained that this had been present for the previous 6–7 years and that he had attributed it to a bee sting to his eye several years earlier.

A chest radiograph showed a large, well-defined mass in the upper half of the right hemithorax, around 18 cm in diameter, associated with

Figure 1 The right-sided Horner syndrome: enophthalmos, partial ptosis and miosis.

displacement of the trachea to the opposite side. Initially, this was thought to be a bronchogenic cyst.

CT (figure 2) helped to further delineate this mass, showing a large, homogeneous, encapsulated lesion in the right upper zone, whose density was mostly similar to fluid, associated with a solid component in the uppermost part. The mediastinal structures, particularly the trachea and right bronchus, were noted to be compressed and displaced to the left side by this lesion. However, no significant spread to adjacent structures, pleural effusion, focal lung lesions or lymphadenopathy were noted.

The mass was excised via a right-sided thoracotomy. At operation, a large, tense mass occupied the right upper thorax, separate from the right upper lobe, which was collapsed (figure 3). The size of the mass precluded surgical excision, so aspiration

Figure 2 CT scan showing a homogeneous lesion in the right upper zone, with compression of the trachea and right bronchus.

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Figure 3 A mass around 15 cm across was completely excised from the right pleural space.

of fluid from the core of the lesion was performed. This yielded about 1 L of blood-stained fluid, thereby reducing the mass to a size that permitted complete surgical excision. The excised mass was a well-circumscribed, cystic structure having a pale brown appearance, measuring 150 mm×152 mm×65 mm (figure 4). On sectioning, it revealed a solid area measuring 120 mm×40 mm×35 mm (figure 5). The rest of the cystic space was lined by a paper-thin lining and filled with serous fluid. Histological examination was consistent with a diagnosis of benign schwannoma with secondary changes of ischaemic necrosis, and a focus of spindle cells with prominent nuclear palisading set in hyalinised stroma (figures 6 and 7). Other solid areas showed proliferation of histiocytes, fibroblasts and haemosiderin deposits. The viable tumour cells were noted to be immunoreactive solely to S-100, a protein found in several tumours, including schwannomas. No malignant changes were evident.

INVESTIGATIONS

- Chest radiograph: large, well-defined mass in the upper half of the right hemithorax, around 18 cm in diameter, associated with displacement of the trachea to the opposite side.
- CT: large, homogeneous, encapsulated lesion in the right upper lobe, whose density was mostly similar to fluid, associated with a solid component in the uppermost part. The mediastinal structures, particularly the trachea and right bronchus, were noted to be compressed and displaced to the left side by this lesion. However, no significant spread to

Figure 4 The right-sided thoracotomy revealing a large mass in the right upper zone. Suction catheter in situ—c. 1 L of blood-stained fluid was drained.

Figure 5 Macroscopic appearance of resected schwannoma.

adjacent structures, pleural effusion, focal lung lesions or lymphadenopathy were noted.

DIFFERENTIAL DIAGNOSIS

- Bronchogenic cyst
- Lung malignancy

OUTCOME AND FOLLOW-UP

Following surgical excision, the patient made an uneventful recovery, with immediate resolution of the shortness of breath and cough, but the HS remained present. He was discharged 5 days postoperatively.

A 3-month follow-up showed that our patient had made a full recovery, with no further asthmatic exacerbations reported.

DISCUSSION

This patient manifested the rare association of a benign intrathoracic schwannoma presenting with HS. The uniqueness of this case lies in the fact that, although HS is a well-known association of sympathetic chain schwannomas in the neck^{2 3} and of malignant apical lung tumours,⁴ it is very uncommon for it to be the presenting feature of a benign intrathoracic lesion.

A literature review was performed via PubMed, using the MeSH terms 'Benign Neoplasms' and 'Horner Syndrome', to identify any similar cases that had been described. This search yielded 12 cases of HS apparently caused by a benign intrathoracic neoplasm. Histologically, a plethora of different lesions have been described, including a chordoma,⁵ thymic cyst,⁶ hydatid

Figure 6 Schwannoma—Antoni A area. Hypercellular areas from the tumour with a proliferation of spindle cells having numerous areas showing prominent nuclear palisading (Verocay bodies; a low power view).

Figure 7 Schwannoma—Antoni B area with numerous foamy macrophages (a high power view).

cyst,⁷ bronchogenic cyst,⁸ aneurysmal bone cyst⁹ and seven cases of benign intrathoracic schwannoma causing HS.^{10–16} Of these cases, the nerve of origin was judged to be a sympathetic nerve in three cases,^{10 12 15} the first thoracic nerve in one case¹¹ and an intercostal nerve in another case.¹³ In this case, the enormous size of the tumour made it exceedingly difficult to identify the nerve of origin.

This case illustrates the importance of a thorough investigation of HS, even in the absence of features suggesting a malignant aetiology, such as weight loss, anorexia or haemoptysis. Despite the benign nature of the lesion in this case, and the earlier presentation of the HS, the tumour was allowed to grow to a size which compromised tracheal integrity and complicated surgical excision via thoracotomy. We therefore suggest that HS should be investigated early and thoroughly.

Learning points

- Horner syndrome is an important presentation that should be investigated fully, even in the absence of systemic features suggesting a malignant aetiology.
- A diagnosis of acute asthmatic exacerbation should be questioned if the patient fails to respond to treatment, as what was described as wheezing, in this case, may well have been stridor.

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Contributors EB provided the histological images from the pathology department, and advised accordingly regarding which images would best convey the messages of the corresponding author. She also suggested relevant captions to the histological images. PJG guided the corresponding author on the more medical aspects of the case, suggesting advice on describing the clinical presentation and management. AM helped the corresponding author throughout the writing of this case report, reviewing the script at every stage and suggesting which photos to submit for review.

Competing interests None.

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