CASE REPORT

Osteochondroma of the clavicle causing Horner's syndrome

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SUMMARY

A 17-year-old girl was admitted with a 1-year history of right-sided headaches and unilateral ptosis for 6 months. She was diagnosed with Horner's syndrome. Ultrasound and CT scan demonstrated an osteochondroma of the posterosuperior aspect of the medial clavicle compressing adjacent structures, including the right internal jugular vein and right common carotid artery. Cartilage sequence MRI showed an osteochondroma with a 5 mm cartilage cap. There were no suspicious features of malignancy. The patient was referred to a specialist centre for surgical resection. Osteochondroma of the clavicle presenting with Horner's syndrome is extremely rare, with only one previously reported case. In this case report, we discuss the anatomy of the oculosympathetic chain, which consists of a three neuron arc. The disruption of the neuronal arc, at any point, may result in Horner's syndrome. We also discuss the various causes of Horner's syndrome and its radiological investigation.

BACKGROUND

Horner's syndrome results from the disruption of sympathetic innervation to the ipsilateral eye (oculosympathetic palsy) and face. The clinical findings of the condition include unilateral ptosis, pupillary meiosis, facial anhidrosis and apparent enophthalmos.¹⁻³ Horner's syndrome may pose a challenge for the clinician in terms of localising the lesion, investigation and treatment. Localising the lesion is potentially difficult due to the complex anatomy of the sympathetic innervation to the eye. Oculosympathetic innervation of the eye ([figure 1](#)) begins with first order (central) neurons that course from the dorsolateral portion of the hypothalamus to the ciliospinal centre of Budge (C8-T2). In the present study they synapse with second-order (preganglionic) neurons, which exit the spinal cord at the level of T1 and pass over the apex of the lung before entering the cervical sympathetic chain in the neck. These second-order neurons synapse in the superior cervical ganglion with third-order (post-ganglionic) neurons at the level of the carotid bifurcation. A branch of third-order neurons innervates the facial sweat glands. The remaining third order neurons follow the internal carotid artery through the cavernous sinus, to innervate the eye. The oculosympathetic neurons innervate the Müller muscle (eyelid retractor) and dilator pupillae muscle ([figure 1](#)).^{1 2} The symptoms, signs and causes of Horner's syndrome therefore vary with the location of the pathology along this complex pathway.

We report a rare case of osteochondroma of the clavicle causing Horner's syndrome. The disruption occurred at the second-order neuron due to the osteochondroma compressing structures on the right side of the neck and disrupting the oculosympathetic neurons at this level. This case is important because it demonstrates a rare cause of Horner's syndrome. Osteochondroma of the clavicle itself is extremely rare, with only a few previously published cases.^{4 5} We have found only one other case in the published literature, in which an osteochondroma of the clavicle caused Horner's syndrome.⁴

Radiological investigations of Horner's syndrome can also be challenging. In the present case, the chest radiograph was normal, most likely because the osteochondroma was hidden from view on the posterosuperior aspect of the medial right clavicle. The patient then had MRI, which was reported as showing a mass at the medial end of the right clavicle, and ultrasound-guided biopsy was recommended ([figure 2](#)). It was only at ultrasound that the osteochondroma was identified ([figure 3](#)). This was then confirmed at CT ([figures 4 and 5](#)). This case is therefore important as it shows the potential difficulty of identifying osteochondroma on different imaging modalities. We discuss the various radiological investigations used in the diagnosis of osteochondroma of the clavicle.

The potential for malignant transformation of solitary osteochondroma is approximately 1%.⁶ In the case published by Kapoor *et al*,⁴ the osteochondroma of the clavicle causing Horner's syndrome underwent malignant transformation into a chondrosarcoma. This potential for malignant transformation demonstrates the importance of identifying and investigating these lesions.

Our case is important in that it adds another lesion that may cause Horner's syndrome to the current published lists. We also discuss the radiological investigation of this condition, as well as the anatomy of the oculosympathetic chain and potential sites where this may be disrupted by various conditions to cause Horner's syndrome.

CASE PRESENTATION

A 17-year-old girl was admitted to the emergency department on 20 March 2015. She presented with a 6-month history of right-sided ptosis. Her symptoms worsened occasionally and were associated with constant watering of the right eye, regardless of the presence of a headache. She reported a 1-year history of right-sided 'pressure-like' headaches with nausea that could last an entire day. The headaches initially occurred three times per week,

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Figure 1 Schematic diagram of the oculosympathetic supply to the eye. V1, ophthalmic nerve; V2, maxillary nerve; V3, mandibular nerve; N, nerve.

but became constant. She was slightly photophobic and phonophobic with the headaches. She occasionally saw a bright flash in her vision on the right side, which lasted up to 5 min. She had experienced three episodes of rhinorrhoea in the past 2 months. She had not noticed any lack of sweating on the right side of the face. She had no significant medical history. She occasionally took ibuprofen (which did not resolve the headaches) and was on Depo (as a contraceptive method). She smoked 10 cigarettes/day and did not drink. Her maternal grandmother suffered from migraines.

On clinical examination, the patient had a meiosis in the right eye, with a pupil diameter of <2 mm, compared to a 5 mm left pupil diameter. She had reduced visual acuity of 9/6 on the right and 7.5/6 on the left. She felt that the right temporal region was mildly reduced on visual field testing. She had full range of eye movements, no evidence of nystagmus and normal corneal reflexes. There was a reduction in sensation in the V1 distribution of the face to pin prick and light touch. There was no facial weakness. The tongue and palate were normal. There were no palpable cervical lymph nodes. Sensation to pin prick was reduced in the right arm in a sleeve-like distribution up to the shoulder, the lower border was at around T4. Vibration and temperature sensation in the arm were normal, as was coordination. No lower limb neurological abnormalities were identified.

Figure 2 MRI (axial T1) demonstrating osteochondroma at the medial end of the right clavicle.

The patient was afebrile. A clinical diagnosis of Horner's syndrome was made.

INVESTIGATIONS

- 30 March 2015: Plain chest radiograph was normal
- 30 March 2015: Blood test results were essentially normal

Full blood count

- White cell count: 4.1 ($4.5\text{--}13.0 \times 10^9/\text{L}$)
- Red blood cell count: 4.56 ($4.10\text{--}5.10 \times 10^{12}/\text{L}$)
- Haemoglobin: 138 (115–160 g/L)
- Haematocrit: 0.41 (0.36–0.46)
- Mean cell volume: 89.3 (80.0–100.0 fL)
- Mean cell haemoglobin: 30.3 (27.0–32.0 pg)
- Mean cell haemoglobin concentration: 339 (310–350 g/L)
- Platelets: 358 ($150\text{--}450 \times 10^9/\text{L}$)
- Neutrophil antibodies: 1.60 ($1.80\text{--}8.00 \times 10^9/\text{L}$)
- Lymphocyte antibodies: 1.93 ($1.10\text{--}6.50 \times 10^9/\text{L}$)
- Monocyte antibodies: 0.34 ($0.20\text{--}0.80 \times 10^9/\text{L}$)
- Eosinophil antibodies: 0.20 ($<0.51 \times 10^9/\text{L}$)
- Basophil antibodies: 0.03 ($<0.11 \times 10^9/\text{L}$)

Figure 3 Ultrasound showing osteochondroma at the medial end of the right clavicle.

Figure 4 CT scan (bone windows) showing an osteochondroma extending posteriorly from the medial end of the right clavicle.

Immunoglobulins

- IgG: 7.93 (6.00–16.00 g/L)
- IgA: 0.77 (0.80–2.80 g/L)
- IgM: 0.96 (0.50–1.90 g/L)

Clotting screen

- Prothrombin time: 10.3 (9.5–13.0 s)
- International normalisation ratio: 1.0
- Activated partial thromboplastin time: 24.3 (21.0–33.0 s)
- Fibrinogen: 3.9 (1.8–4.0 g/L)

Renal electrolytes

- Sodium: 142 (133–146 mmol/L)
- Potassium: 4.2 (3.5–5.3 mmol/L)
- Bicarbonate: 24 (22–29 mmol/L)
- Urea: 2.7 (2.5–7.8 mmol/L)
- Creatinine: 49 (50–95 µmol/L)

Liver function tests

- Bilirubin: 6 (<21 µmol/L)
- Alkaline phosphatase: 106 (34–150 U/L)
- Alanine aminotransferase: 18 (5–40 U/L)
- Albumin: 43 (35–50 g/L)

Complement

- C3: 1.30 (0.80–2.10 g/L)
- C4: 0.32 (0.15–0.50 g/L)

Bone profile

- Alkaline phosphatase: 106 (34–150 U/L)
- Albumin: 43 (35–50 g/L)
- Calcium: 2.45 (2.2–2.6 mmol/L)
- Calcium (adjusted): 2.43 (2.2–2.6 mmol/L)
- Plasma viscosity: 1.69 (1.50–1.72 mPa s)
- Vitamin B₁₂: 280 (180–900 pg/mL)
- Serum folate: 3.0 (4.4–20.0 µg/L)
- Serum ferritin: 38 (29–470 µg/L)
- C reactive protein: 2 (<6 mg/L)
- Thyroid-stimulating hormone: 3.3 (0.27–4.20 mU/L)
- Total protein: 75 (60–80 g/L)

Figure 5 CT scan (soft tissue windows) showing an osteochondroma extending posteriorly from the medial end of the right clavicle, compressing adjacent structures in the right side of the neck, including the right internal jugular vein and right common carotid artery.

Figure 6 MRI (coronal short τ inversion recovery) showing osteochondroma of the posterosuperior aspect of the medial right clavicle.

- Glycated haemoglobin: 34 (mmol/mol)
- Free T4: 14.1 (12.0–22.0 pmol/L)
- Creatine kinase: 70 (25–200 U/L)
- Rheumatoid factor: 4 (<20 IU/mL)
- ACE inhibitors: 48 (8–52 IU/L)
- Antithyroid peroxidase: <20 (<100 IU/mL)

- 16 April 2015: MRI of the head pre-gadolinium and post-gadolinium, neck MR angiogram and neck MRI with gadolinium were carried out. The report of the neck MRI described a large heterogeneous mass, which was low signal intensity on T1 (figure 2) and relatively hyperintense on T2-weighted imaging, with areas of enhancement peripherally, in the right supraclavicular fossa. This was accompanied by a surrounding high T2 signal indicative of inflammatory fluid. The lesion was abutting the lateral aspect of the proximal right common carotid artery, but the artery was seen to be patent. The mass was distinct from the thyroid gland and was not in the lung apex. No intracranial abnormalities were detected. A necrotic lymph node was suggested as the most

Table 1 Potential causes of Horner's syndrome

First order (central)	Second order (preganglionic)	Third order (postganglionic)
Hypothalamus	Lung	Superior cervical ganglion
► Stroke	► Apical (Pancoast) tumour	► Trauma
► Tumour	Clavicle	► Tumour in adjacent structures
Brainstem	► Trauma	► Jugular venous ectasia
► Stroke	► Tumour	► Iatrogenic (surgical neck dissection)
► Tumour	Iatrogenic	Internal carotid artery
► Demyelination	► Jugular cannulation	► Trauma
Cervicothoracic spinal cord	► Thoracic surgery	► Dissection
► Trauma	Cervical rib	► Aneurysm
► Tumour	Subclavian artery aneurysm	► Arteritis
► Demyelination	Thyroid tumour	Skull base
► Syringomyelia	Brachial plexus injury	► Tumour (pituitary)
► Myelitis		► Inflammation
		► Thrombosis
		Cluster headache

The causes for the first-order neuron Horner's syndrome include cerebral vascular accident (stroke), lateral medullary infarct, trauma, tumour and demyelinating disease. Second-order neuron causes include Pancoast tumour, vascular aneurysm and thyroid tumour. Third-order neuron causes include internal carotid artery dissection or aneurysm, and skull base tumour. The differential diagnoses for the clavicular mass include lymphadenopathy, abscess, malignancy, arteriovenous malformation and vessel aneurysm.

likely cause of the symptoms and ultrasound-guided biopsy was recommended.

- ▶ 8 May 2015: Ultrasound demonstrated a bony mass extending from the medial clavicular head in the right supraclavicular fossa, suggestive of an osteochondroma (figure 3).
- ▶ 8 May 2015: CT of the thorax confirmed a large osteochondroma growing from the posterosuperior aspect of the medial clavicle and causing soft tissue impingement (figures 4 and 5).
- ▶ 28 May 2015: Repeat MRI using cartilage-sensitive sequences demonstrated an osteochondroma of approximately 4×3 cm at the posterosuperior aspect of the distal end of the clavicle (figure 6). The cartilage cap measured approximately 5 mm. The osteochondroma was causing displacement of the soft tissues at the root of the neck, including the right internal jugular vein and right common carotid artery. No malignant transformation was identified.

DIFFERENTIAL DIAGNOSIS

Differential diagnoses and causes of Horner's syndrome are summarised in table 1.

TREATMENT

The patient has been followed up in clinic and has been referred to a specialist centre for surgical resection.

OUTCOME AND FOLLOW-UP

At 3 months following initial presentation, there has been no significant change to the patient's condition and she is awaiting surgical excision.

DISCUSSION

Horner's syndrome is caused by disruption of the sympathetic innervation to the eye (oculosympathetic palsy) and face at any part of the sympathetic arc from the hypothalamus to the eye and face. This is a complex arc made up of a three-neuron chain, as illustrated in figure 1. The first-order neurons arise from the dorsolateral hypothalamus and descend through the brainstem to the ciliospinal centre of Budge, which is located between C8 and T2 of the cervical spinal cord. At this point, the neurons form a synaptic link with second-order neurons, which exit the anterior horn of the spinal cord. These neurons follow a path over the lung apex to join the sympathetic chain in the neck, where they form synapses in the superior cervical ganglion. It is between exiting the ciliospinal centre of Budge and entering the superior cervical ganglion in the neck that the oculosympathetic chain was disrupted in the case that we present, resulting in second-order Horner's syndrome. This caused a disconnect between the second-order and third-order neurons. The cell bodies of third-order neurons give rise to postganglionic axons that travel alongside the internal carotid artery, eventually dividing to supply the Müller muscle (eyelid retractor) and dilator pupillae muscle.^{1 2}

Knowledge of the oculosympathetic supply to the eye and its pathway is essential in order to locate the site of lesions that may cause its disruption and result in Horner's syndrome. It is also important for the clinician to be aware of the varying causes of Horner's syndrome at the different parts of the neuronal arc. These causes include cerebral vascular accident (stroke), lateral medullary infarct, trauma, tumour and demyelinating disease causing first-order neuron disruption. Pancoast tumour, vascular aneurysm and thyroid tumour cause second-order disruption. Internal carotid artery dissection, internal carotid artery aneurysm and skull base tumours cause

third-order neuron disruption (table 1). We have found only one previous case of second-order Horner's syndrome resulting from osteochondroma of the clavicle.⁴ This was a case of a 28-year-old man with a 3-year history of left-sided shoulder pain and ptosis. CT scan confirmed a diagnosis of osteochondroma of the medial left clavicle. The mass was excised and histology demonstrated a low-grade chondrosarcoma. At 1-year follow-up, the patient had no recurrence or pain, but his ptosis was still present. The risk of malignant transformation of solitary osteochondroma is approximately 1%.⁶ Although this is relatively low, the case by Kapoor *et al*⁴ illustrates the importance of radiological investigations or excision biopsy for histological analysis. It is important for the radiologist to be familiar with the imaging features of osteochondroma; as our case illustrates, the patient underwent MRI following a normal chest radiograph, and the MRI appearances were reported as showing a mass adjacent to the clavicle and a biopsy was recommended. It was only at the time of the planned biopsy at ultrasound that the osteochondroma was identified, then confirmed at CT.

In this case report, we have added osteochondroma of the clavicle as yet another cause of Horner's syndrome. We have discussed the anatomy of the oculosympathetic pathway and areas where it may be disrupted, leading to Horner's syndrome. Methods of radiological investigation, including some of the difficulties involved, have also been highlighted.

Learning points

- ▶ Osteochondroma of the clavicle, although rare, should be considered in the differentials for causes of Horner's syndrome.
- ▶ Radiologists should be aware of the potential for misdiagnosis of an osteochondroma of the clavicle as a soft tissue mass. If in doubt, they should carry out an ultrasound and/or CT scan, which would better demonstrate the bony anatomy and therefore more easily diagnose osteochondroma.
- ▶ Osteochondroma of the posterosuperior aspect of the clavicle may not be visible on plain radiograph, and therefore other imaging modalities such as ultrasound and/or CT scan may be necessary to visualise the osteochondroma.
- ▶ The incidence of malignant transformation in solitary osteochondroma is approximately 1%. Therefore, although the incidence of malignancy is low, it is important to identify these lesions and perform the appropriate tests to confirm that the lesion is benign.

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