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Spontaneous cervical haematoma resulting from intrathoracic pathology

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Abstract

Spontaneous cervical haematomas are rare occurrences for which a wide variety of aetiologies have been reported. Although the pathogenesis of this condition can be quite diverse, all known cases have emanated from a cervical source. The authors report the first case of a spontaneous cervical haematoma arising from an intrathoracic source. A case review and Medline search from 1962–2003 was carried out.

No prior experience with intrathoracic sources for cervical haematomas has previously been reported. Transcervical haematoma evacuation confirmed the intrathoracic origin and a median sternotomy was required to gain control of this haemorrhage.

Thoracic sources should be considered in the differential diagnosis of a spontaneous cervical haematoma. Preparations for a combined procedure with a thoracic surgeon can be predicted pre-operatively based on imaging studies.

Key words: Neck; Haematoma; Airway Obstruction; Thoracic Surgical Procedures

Introduction

Spontaneous cervical haematomas are rare entities associated with a myriad unusual aetiologies. Among the panoply of conditions associated with spontaneous cervical haematomas are violent sneezing,[1] anticoagulation,[1,2] polycythaemia rubra vera,[1,3] parathyroid adenoma,[4,5] trauma,[6] ingested foreign bodies,[7] neck infections,[8] and connective tissue disorders.[9,10]

Spontaneous thoracic haematomas are correspondingly rare lesions, virtually unreported in the Otolaryngology literature; however, they often involve patients with similar risk factors, especially those with connective tissue disorders. In reviewing the existing literature surrounding spontaneous haematomas of the neck and chest the authors have not located a single case of thoracic disease presenting with clinical findings limited to the neck. They present here the first report of a case of spontaneous cervical haematoma found to have an entirely intrathoracic source.

Case report

A 33-year-old white female with a history of NF1 and Marfan's syndrome presented to the Emergency Department (ED) of this institution with the chief complaint of progressive swelling of the right side of her neck. The patient had been in her usual state of health, denied any recent infections, trauma, or participation in activities that involved manipulation of the neck. The lower portion of the right side of her neck continued to increase in size while she was observed in the ED and the Otolaryngology service was consulted.

The patient was found to have a firm mass superior to the right clavicular head without evidence of pulsations or audible bruits. The overlying skin was normal in appearance and the neck and chest were free of surgical scars. The patient had no stridor nor voice changes and, despite some pain and a heavy sensation in her neck, the airway was considered stable.

Coagulation studies and complete blood count were within normal limits. The patient's vital signs were stable (with a sinus tachycardia of 100/minute), fluid resuscitation was initiated and the patient was considered safe to undergo computer tomography (CT) scan and angiography imaging studies.

CT scan identified a $13 \times 7 \times 4$ cm intermediate attenuation mass extending from the level of the hyoid bone inferiorly to the level of the sternomanubrial junction mildly compressing the middle portion of the trachea (Figure 1). The right clavicular head and sternoclavicular junction exhibited bony changes consistent with a chronic process (Figure 2). The close relationship between the chronic bony changes at the right clavicular head and the mass lesion were worrying in case the acute haemorrhage was from a pre-existing neoplasm. An arteriogram demonstrated normal anatomy of the aortic arch and cervical area without evidence of contrast extravasation, aneurismal dilation, ulcerations or stenoses. The possibility of an intrathoracic haemorrhage in a patient with Marfan syndrome and NF1 led the authors to consult the cardiothoracic surgery service for assistance in managing a disease process extending beyond the anatomical territory of the head and neck surgeon.

The continued expansion of this haematoma in conjunction with the authors' failure to identify a bleeding source in the neck or chest with CT scan or angiogram made

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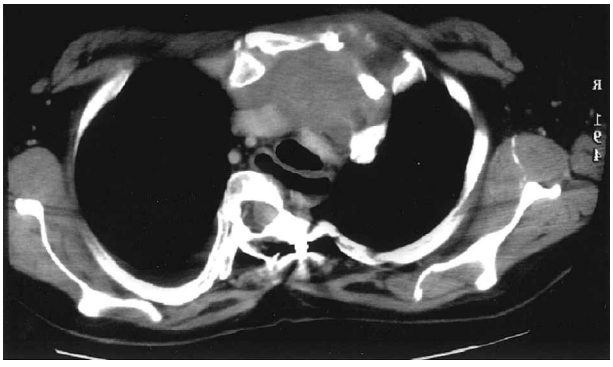

FIG. 1
Cervical haematoma at the level of the glottis.

- **Spontaneous cervical haematomas are rare and all previously reported cases have resulted from pathology in the cervical region**
- **In this case the haematoma was seemingly due to intrathoracic bleeding**
- **The authors report this as an unique case and have validated this with a Medline search**

surgical exploration the only viable treatment option. The patient was taken to the operating theatre for a neck exploration, but was jointly consented by the cardiothoracic surgery service for the possibility of a thoracic operation to identify and treat the source of this haemorrhage.

Exploration of the right side of the neck was undertaken through a standard apron incision. Upon dividing the platysma, a large haematoma tracking beneath the inferior skin flap at least to the level of the clavicle was encountered and removed. The right carotid sheath was inspected and found to be normal. The authors failed to identify the source of bleeding in the neck, yet blood continued to flow briskly from the tissue located at the distorted right clavicular head. Progressive removal of the tissue in this location produced further bleeding that was refractory both to electrocautery and packing with Surgicel. The authors continued to remove tissue believed to be an organized clot found adjacent to the clavicular head, and sent several portions to pathology for permanent section.

The cardiothoracic surgeon evaluated this persistent bleeding emanating from a location deep to the sternum and elected to proceed with a median sternotomy. After obtaining adequate exposure, evacuating a retrosternal haematoma and controlling the bleeding associated with opening the chest, a single vascular injury was identified. An avulsed perforating branch of the right IMA was located at the second interspace and was treated with suture ligation of the branch as well as application of vascular clips to the IMA itself.

The patient's post-operative course was unremarkable. She was extubated on POD#1 and transferred to a general nursing floor on POD#2. She was discharged to home on POD#6 and has been followed for over 15 months without any further bleeding episodes.

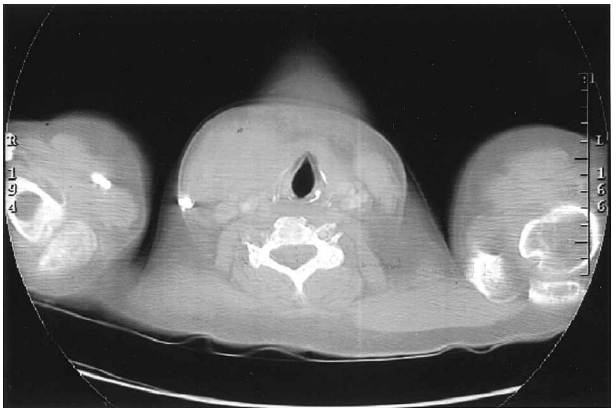

FIG. 2
Right clavicular head erosion and intrathoracic extension of haematoma. Note Harrington spinal rods used in previous scoliosis repair.

Pathologic examination of the substernal tissue was reported as diffuse neurofibroma with extensive fresh haemorrhage, having readily identifiable Wagner-Meissner bodies, without evidence of atypia or malignant transformation.

Discussion

Neurofibromatosis type 1 (NF1), or von Recklinghausen's disease, is an autosomal dominant disorder of variable penetrance found in approximately 1 in 3000 Americans.[10] The diverse clinical manifestations of this disease are a result of both mesodermal and ectodermal dysplasia, most commonly manifest in café-au-lait spots, neurofibromas, gliomas, Lisch nodules (pigmented iris hamartomas), and meningeal lesions.[10,11,12] Although not a common feature of neurofibromas, haemorrhage occurring either spontaneously or after minimal trauma is a potentially lethal complication of this disease. A retrospective review of plexiform neurofibromas confirmed the rarity of these bleeding events when it estimated an incidence of 0.005 spontaneous haematomas per patient with a plexiform neurofibroma per year of follow-up.[13]

In addition to the risk of haemorrhage from within a neurofibroma, patients are also thought to have the possibility of vessel wall abnormalities resulting from the contribution of neural crest cells to the adventitia and smooth muscle of blood vessels.[11] Arterial dysplasia in NF1 (vascular neurofibromatosis) can be identified during histological examination of an involved vessel, and may be present in over three per cent of NF1 patients.[10,11] Arterial dysplasia may explain the increased vascular fragility experienced during surgical procedures as well as the aetiology of spontaneous vessel rupture in the NF1 patient population.[10]

The patient presented in this case report had an expanding cervical haematoma that almost certainly resulted from acute haemorrhage in a long-standing intrathoracic neurofibroma. Erosive changes in the sternum and right sternoclavicular joint are indicative of a chronic process, and the pathologist confirmed the presence of diffuse neurofibroma tissue within the specimen removed from the substernal location. The identification of an avulsed branch of the right IMA raises the interesting possibility of arterial dysplasia as a second, or alternate, source of bleeding; however, this possibility is quite remote. It is most likely that the vessel avulsion occurred as a result of operative trauma sustained during median sternotomy and was not responsible for the

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patient's dramatic initial presentation. Biopsy of the avulsed IMA branch with histopathological identification of mucoid substance and proliferation of myointimal cells in the arterial intima would support a role for vascular neurofibromatosis in predisposing the vessel to operative trauma, yet vessel biopsy was not performed.[10]

Conclusion

Spontaneous cervical haematoma is a rare condition with a variety of reported aetiologies. The authors report here the first case of an intrathoracic process resulting in a spontaneous cervical haematoma. While the head and neck surgeon may be comfortable with the management of cervical haematomas, an intrathoracic source of bleeding could present significant difficulties in operative management. When evaluating similar cases of cervical haematoma the otolaryngologist should have a higher index of suspicion for unusual locations of tumour haemorrhage or vessel disruption, especially in patients with connective tissue disorders.

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