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Limited form of Churg-Strauss syndrome presenting as a mass in the neck

J. W. Moor, M.R.C.S., J. U-King Im, M.R.C.S.*, A. W. MacDonald, F.R.C.Path.†, E. Whitehead, F.R.C.S.‡

Abstract
A 53-year-old man was referred to the ENT department with a large mass in the left supraclavicular fossa. The histological diagnosis showed the mass to have arisen due to a granulomatous vasculitis consistent with Churg-Strauss syndrome (CSS). CSS usually comprises asthma, eosinophilia and systemic vasculitis although limited forms of the disease exist where one of these diagnostic criteria is missing. This is one such case as the patient was non-asthmatic.

Key words: Churg Strauss Syndrome; Neck; Neck Neoplasms

Introduction

Churg-Strauss syndrome (CSS) is a systemic vasculitis characterized by asthma, raised peripheral blood eosinophilia and granulomatous vasculitis.[1] This was initially described in 1951 following a pathological study of patients who had suffered from asthma, vasculitis affecting various organs, peripheral blood eosinophilia, and died from the disease.[2] Since then various cases have been reported where one of the three main diagnostic criteria (above) has been missing. These are limited forms of CSS.[3,4]

We report one such case of a man presenting with a neck lump who was diagnosed histologically as having CSS, without preceding asthma.

Case report

A 53-year-old man was referred to the ENT department by his General Surgery Consultant who had been treating the patient for long-standing ulcerative colitis. Five years earlier he had undergone proctocolectomy and ileostomy for Dukes A adenocarcinoma discovered on routine colonoscopic monitoring of this disease. Despite the original histological stage of Dukes A adenocarcinoma, it was felt that the chances of the mass representing malignant lymphadenopathy were very high.

The mass had arisen over a matter of several weeks, it was non-tender.

In the past he had suffered with nasal polyposis requiring nasal polypectomy on two occasions. For many years he had a persistent low grade proteinuria. He had suffered from ulcerative colitis for nearly 20 years and had never suffered from asthma.

On examination he had a firm mass arising at the lower border of the clavicular head of sternocleidomastoid, approximately 5 cm diameter. It was not attached to the skin but was fixed to deeper layers. There was no overlying erythema and no other lymphadenopathy.

A full blood count showed a raised eosinophil count of 0.46×10^9/l, equivalent to 10 per cent of the white blood cell count, and a chest X-ray showed that the heart size was normal and that the lung fields contained no focal or diffuse abnormalities.

A Tru-cut biopsy was undertaken. The tissue sample did not show any evidence of neoplasia; it was composed of loose fibrous connective tissue containing blood vessels and inflammatory cells, mainly composed of lymphocytes and eosinophils. The exact diagnosis was unclear from this specimen. Prior to further tissue sampling an magnetic resonance image (MRI) scan was performed.

This showed a 3 cm mass in the left route of the neck just above the clavicle, related to the posterior border of the sternomastoid and possibly involving the muscle (Figure 1(a) and (b)).

A formal open biopsy was undertaken and following inspection by Dr A. MacDonald (co-author), the slides were reviewed by a soft tissue tumour expert at the Soft Tissue Tumour Unit, Guys and St Thomas's Hospital, London. They showed heavily inflamed tissues, with a florid and focally occlusive granulomatous vasculitis with a very large number of eosinophils (Figures 2 and 3). Histologically this represented Churg-Strauss syndrome.

Autoimmune profile and c-ANCA studies were performed, both of which were negative.

Initially he remained stable and received no specific treatment, he was referred to the respiratory physicians for further care. Over the next two months the eosinophil count rose to 14 per cent of the total white cell count. Slowly the mass enlarged and required treatment, oral methylprednisolone was commenced, initially 20 mg daily. The mass reduced in size and eventually disappeared. The eosinophil count returned to normal levels. Over the following three years the dose of steroid was reduced and stopped with no recurrence of the mass, and no onset of respiratory, cardiac or other systemic problems.

To our knowledge the patient has been symptom free for five years.

From the Department of ENT Surgery, Leeds General Infirmary, Leeds and the Departments of General Surgery*, Histopathology† and ENT Surgery‡, Hull Royal Infirmary, Hull, UK.
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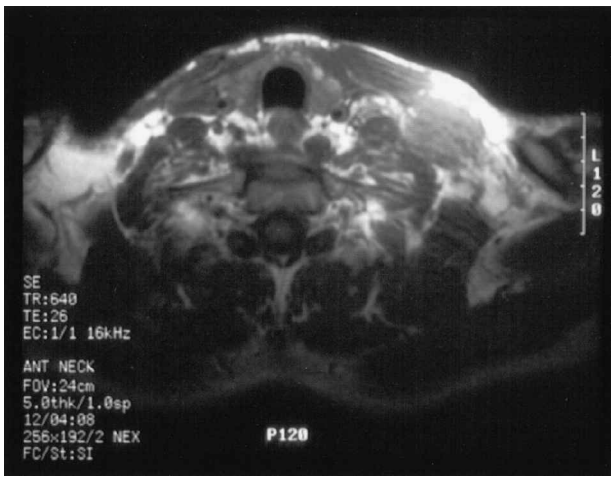
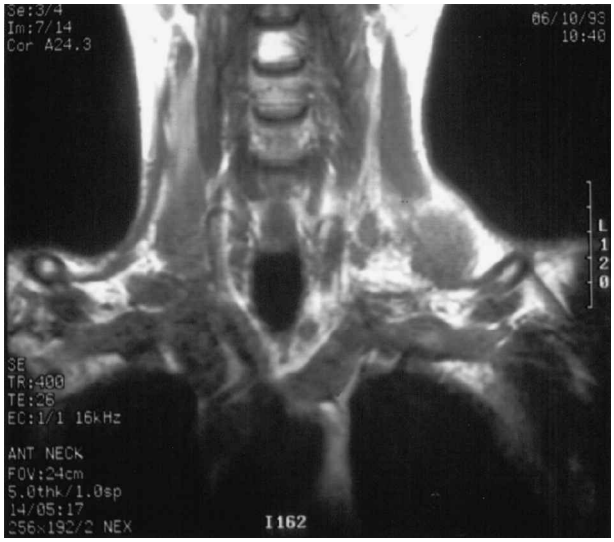


(a)



(b)

FIG. 1

(a) and (b) (MRI scans). The mass is lateral to the great vessels, its inferior aspect immediately adjacent to portions of the brachial plexus.

Discussion

Churg-Strauss syndrome is a rare systemic vasculitis potentially involving pulmonary, cardiac, gastrointestinal, skin, renal and nervous systems.[5,6] The disease is characterized by pulmonary and systemic necrotizing angiitis, extravascular granulomas, and eosinophilic infiltrates occurring almost exclusively in patients with a history of asthma or allergy. It occurs equally in both sexes and can present at any age, the mean age at onset being 40 years. It is recognized that prior to developing vasculitis, there is often a prodrome of increasingly severe asthma, followed by pulmonary or gastrointestinal eosinophilic infiltration.[7] The clinical diagnosis is based on the three criteria of asthma, a peak peripheral blood eosinophil count of $>1.5 \times 10^9$, and systemic vasculitis involving two or more extrapulmonary organs. The limited form of CSS is a pathological lesion that does not fulfil the complete criteria for classic CSS.[4]

There have not been many previous reports of the limited forms of CSS, Lie (1993) reported that only 12 cases had previously been detailed in the literature up until 1991, although after a detailed search of his own consultation files, in 1993 he was able to publish a further 21 other cases.[4] In limited CSS, the gastrointestinal tract is the most commonly affected, other cases show involvement of almost any organ as the primary manifestation of the disease; gall bladder, calf muscle, bladder, coronary artery etc.[4] To our knowledge there have been no previous reports of limited CSS masquerading as metastatic lymphadenopathy in the neck, or in the ORL literature.

Of note in our case is the antecedent history of nasal polyposis, although the exact cause of the nasal polyposis is not confirmed by any histological diagnosis in the clinical notes. Nasal eosinophilic polyposis, crusting of the nasal mucosa and chronic or recurrent eosinophilic sinusitis are common in the evolution of CSS. Also the patient had, over many years, repeatedly undergone dipstick urinalysis, which consistently revealed a low-grade proteinuria. This had never been thoroughly investigated and no diagnosis had been recorded. It may represent a further manifestation of the limited form of CSS; with the patient now in remission it will never be proven.

It is important to recognize both CSS and the limited form of CSS as both can have a high morbidity and mortality[8] as a result of cardiac involvement.[6] Treatment consists of high dose oral corticosteroids to which CSS rapidly responds. If there is an imminent threat to major organs by advancing disease then intravenous pulses of methylprednisolone should be used. Cyclophosphamide can be administered in resistant cases.

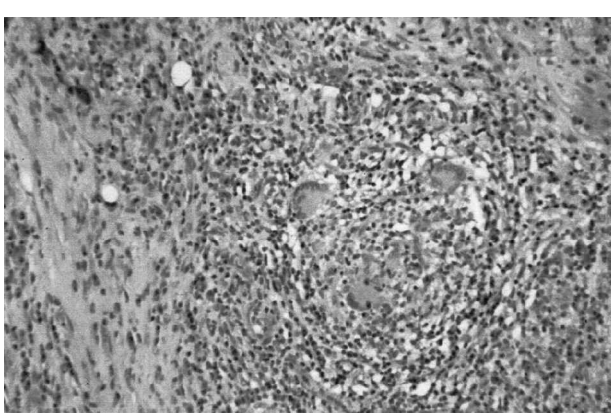


FIG. 2

Granuloma containing giant cells. The slide shows heavy infiltration of surrounding connective tissue by eosinophils (H & E; ×50).

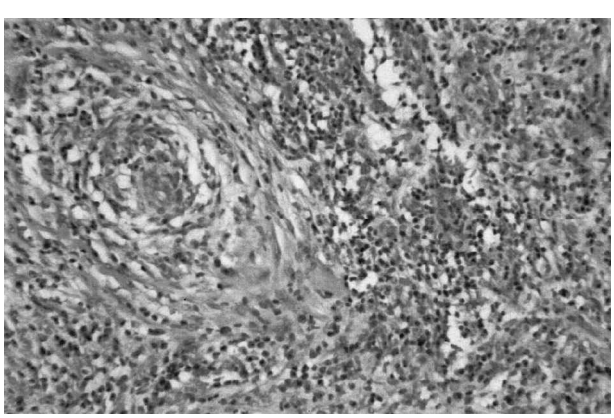


FIG. 3

Blood vessel infiltrated by eosinophils and lymphocytes. The lumen has been obliterated (H & E; ×50)

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It is unlikely that most ENT surgeons will ever encounter a patient with either CSS or the limited form of CSS; the upper aerodigestive tract is rarely affected and there are no other reported cases of CSS presenting as a neck mass. Allergic rhinitis does occur in about 75 per cent[6,9] of patients and nasal eosinophilic polyposis and sinusitis are relatively common. Perforated nasal septa have been reported.[9,10] Whether these clinical entities routinely become sufficiently problematic to warrant specialist referral is unclear.

This case highlights the importance of obtaining an accurate histological diagnosis, even if several attempts are required to achieve this. We recommend that if faced with a patient who has a histological diagnosis of either CSS or the limited form of CSS that the patient is urgently referred to a respiratory physician. Further management of specific ENT-related problems can then be dealt with at a multidisciplinary level.

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Department of Medical Photography, Hull Royal Infirmary.

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Address for correspondence:
Mr E. Whitehead,
Department of ENT Surgery,
Hull Royal Infirmary,
Anlaby Road,
Hull HU3 2JZ, UK.

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