disc with eventual progression to ankylosis of the spine. Early inflammatory lesions are common at the vertebral entheses, notably at the junction of the annulus fibrosus with the periosteum of the vertebra. Although not usually visualized on radiographs, these early changes are identifiable on MRI as focal areas of bone oedema.

Spondylodiscitis associated with Crohn’s disease has been reported twice [9, 10]. The association of juvenile-onset SpA with Crohn’s disease may only emerge later in the course of the disease [11]. Spondylodiscitis can be the initial manifestation of AS and lead to diagnostic confusion. This case illustrates the dramatic MRI changes in the presence of minor plain radiographic features at initial diagnosis and the prompt clinical, laboratory and MRI improvement following infliximab therapy.

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Key messages

- Spondylodiscitis can be the initial manifestation of AS and is responsive to anti-TNF-α therapy.

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Epithelioid haemangioendothelioma and paraneoplastic hypercorticisms

Sir. A 24-yr-old man was followed for 13 yr for Cushing’s syndrome. He had two hypophysectomies (the first was partial and the second complete). Histological examination did not confirm an adenoma and the surgery did not bring about a decrease of secretion of adrenocorticotropic hormone (ACTH). The outcome extension was negative. Bilateral adrenal gland surgery did not improve the Cushing’s syndrome nor the ACTH secretion.

In 2002, after a small trauma, the patient presented with violent pain in the right leg, mainly in the ankle. On examination he was found to have oedema of the right calf. The remainder of the physical examination was normal except for the Cushing’s syndrome which was already known. Right ankle radiographs showed a radiolucent area with a well-defined rim of bone. Magnetic resonance imaging (Fig. 1) showed multiple areas of bone involvement with increased intensity on T2-weighted images; there were the same lesions in muscle. Biopsies of the bone and muscle lesions confirmed the diagnosis of epithelioid haemangioendothelioma (HE), characterized by the presence of epithelioid and histolytic endothelial cells with primitive vascular differentiation. The tumor was surgically removed and the patient treated with radiation therapy. Excision of the tumour mass permitted prompt biochemical remission of hypercortisolism and normalization of ACTH secretion for the first time for more than 13 yr. The clinical presentation and biological evolution favoured a diagnosis of paraneoplastic Cushing’s syndrome.

HE is a rare malignant vascular neoplasm described in soft tissue and bone. It seems to have a predilection for males and to affect all age groups, but mainly patients in the second and third decade. The most frequent symptoms are pain and fracture. Multicentric involvement has been reported in 22 to 64% of patients. Paraneoplastic Cushing’s syndrome is also a rare entity; it results from ectopic corticotropin-releasing secretion. In some cases the primary tumour presents late. Some studies have demonstrated the synthesis of corticotrophin-releasing factor in non-neuroendocrine tissues.

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Fig. 1. Ankle RMN: multiple areas of bone involvement.
Two cases of meningeal involvement in Wegener’s granulomatosis

Sir, Meningeal involvement in Wegener’s granulomatosis (WG) has rarely been described [1]. In a systematic English literature review using electronic bibliographic databases (EMBASE, MEDLINE, CINAHL) we found only 43 cases previously reported. Magnetic resonance imaging (MRI) with gadolinium contrast is the most useful technique for detecting meningeal disease. Two distinct patterns of thickening distribution in WG were recognized by Murphy et al. in 1999 [2]: diffusely abnormal meninges unrelated to sinus or orbital disease and focal dural enhancing thickening adjacent to sinus or orbital disease. We have observed two cases of this rare condition:

Case 1 (GP, male): at the age of 49 this patient experienced his first occurrence of chronic crusty-bloody rhinitis and sinusitis, unresponsive to conventional topical therapies. One year later, in 2000, he visited an ophthalmologist for the abrupt onset of double vision and partial visual loss in the right eye. Ophthalmic evaluation revealed a swollen disc and horizontal diplopia. MRI of the brain was normal. Diagnosis of primary retrobulbar optic neuropathy was made and treatment with topical injection of corticosteroids (CS) led to resolution. In the following months the patient experienced worsening of sinonasal symptoms and multiple recurrences of optic neuropathy during tapering of CS therapy. In 2001 the patient came to our attention for severe exacerbation of ocular abnormalities during low-dose therapy. Right eye pain, blurred vision and diffuse headache were present. Fundus examination revealed severe optic disc oedema and high-dose CS treatment had to be restored. A paranasal sinus computed tomography (CT) scan disclosed thickening of maxillary sinus and nasal mucosa and otolaryngologist evaluation described septal perforation and diffuse crusty rhinitis. MRI of the brain showed a diffuse enhancing leptomeningeal thickening over the convexity of the right hemisphere and bilaterally over the interhemispheric fissure and the intracranial surface of sphenoidal greater wings (Fig. 1A). The right mass extended to the superior orbital fissure. Examination of cerebrospinal fluid revealed only a moderate pleocytosis and a chest X-ray was normal. The erythrocyte sedimentation rate (ESR) was 87 mm/h, C-reactive protein (CRP) 27 mg/dl and antinuclear antibodies were 1:320. Antineutrophil cytoplasmic antibodies (ANCA) were absent. A paranasal mucosal biopsy showed necrotizing granulomas and areas of leucocytoclastic small vessel vasculitis. Also a meningeal biopsy was performed and a T-lymphocytic inflammatory picture was found. Diagnosis of WG was made according to 1990 ACR criteria [3] and treatment with oral cyclophosphamide 150 mg, prednisone 1 mg/kg body weight and cotrimoxazole was started. In few weeks the patient’s symptoms completely resolved and inflammatory indices returned to normal. After 15 months immunosuppressive therapy was shifted to oral methotrexate, 20 mg/week. Following imaging revealed no residual meningeal disease.

FIG. 1. T₁-weighted, contrast-enhanced MRI scan of the head. (A) Case 1: diffuse leptomeningeal thickening over the convexity of the right hemisphere and bilaterally over the interhemispheric fissure. (B) Case 2: thickening of left-sided leptomeninges and dura of the skull base.