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Clinical Records

Relapsing polychondritis associated with monoclonal gammopathy in a patient with myelodysplastic syndrome

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Abstract
Relapsing polychondritis is a rare condition characterized by inflammation and subsequent degeneration of cartilages. Deformity of the pinna, nasal saddling and stridor due to involvement of the cartilages of the respiratory tract may lead to patients being referred to the otolaryngologist for initial assessment and further management. Recent observations have suggested that relapsing polychondritis may occur as a paraneoplastic phenomenon in cases of myelodysplasia. The case of a patient with relapsing polychondritis, myelodysplastic syndrome and a monoclonal gammopathy is presented. The authors highlight the apparent existence of this association and encourage otolaryngologists to consider such possible links when cases of relapsing polychondritis present to the outpatients department.

Key words: Polychondritis, Relapsing; Myelodysplastic Syndromes; Monoclonal Gammopathy, Benign

Case report

In 1995 a 65-year-old retired bricklayer presented to his general practitioner with weight loss of six kilograms in one year. He had chronic obstructive pulmonary disease (COPD) and osteoarthritis of the hands and hips. He had undergone left total hip replacement in 1994. At presentation his haemoglobin was 134 g/l, white cell count 11.6×10^9/l, platelets 267×10^9/l and monocytes 3.0×10^9/l (normal less than 0.9×10^9/l). The blood film showed monocytosis and hypogranular neutrophils. A bone marrow aspirate showed granulocytic hyperplasia and monocytosis with minimal dysplasia. Stainable iron was present in macrophages. Biopsy of a palpable axillary lymph node showed inflammation and fibrosis only. His weight loss was attributed to worsening COPD. His medical records showed a monocytosis of 1.4×10^9/l in 1986 when he was first investigated for arthritis.

In 1996 he underwent right total hip replacement. Following this he was anaemic with a haemoglobin of 102 g/l and mean cell volume (MCV) of 78.7 fl, and he was treated with oral iron.

Over the next year the patient's weight and blood count remained stable. His haemoglobin was 136 g/l, white cell count 13.9×10^9/l monocytes 4.6×10^9/l and platelets 326×10^9/l. No treatment was given. In 1997 the patient noticed a gradual but progressive deformity of both pinnae. He complained of muffled hearing, intermittent hoarseness and morning stiffness affecting the neck, shoulders and hands. He was noted to have hepatomegaly. Liver function tests were normal. His white cell count had increased to 25.8×10^9/l with a monocyte count of 8.2×10^9/l. Haemoglobin was 136 g/l and platelets 277×10^9/l. A monoclonal IgM kappa band was noted; total IgM 3.96 g/l (normal 0.57–2.66 g/l). Other immunoglobulins were normal. Repeat bone marrow aspirate showed granulocytic hyperplasia, dysplastic granulocytes and monocytosis. Blast cells were less than 10 per cent of nucleated cells. Chromosomal studies from this bone marrow aspirate showed a normal male karyotype. A range of auto-antibody tests was negative. Computerized tomography (CT) scan of the chest and abdomen confirmed gross hepatomegaly with no focal lesion and no lymphadenopathy or splenomegaly. Magnetic resonance imaging (MRI) of the laryngeal and tracheal cartilages could not be performed due to the patient's severe breathlessness secondary to COPD.

He was referred to the department of otolaryngology for assessment of his deformed pinnae and muffled hearing. He was noted to have thickening and deformation of the auricle and collapse of the cartilaginous external auditory canals (Figure 1). Pure tone audiogram showed symmetrical high frequency sensorineural hearing loss consistent with his age, with a super added conductive deficit from canal wall collapse. Ear moulds which stented the external auditory meatuses were made in the audiology department. These improved the patient's hearing to a satisfactory level.

He was referred to the rheumatology department, which supported the diagnosis of relapsing polychondritis in the absence of any other rheumatological associations. Pinna biopsy was requested and showed focal degeneration of cartilage with mild inflammation in keeping with the clinical picture. Intermittent hoarseness was thought to be due to inflammation of the laryngeal cartilages. There was

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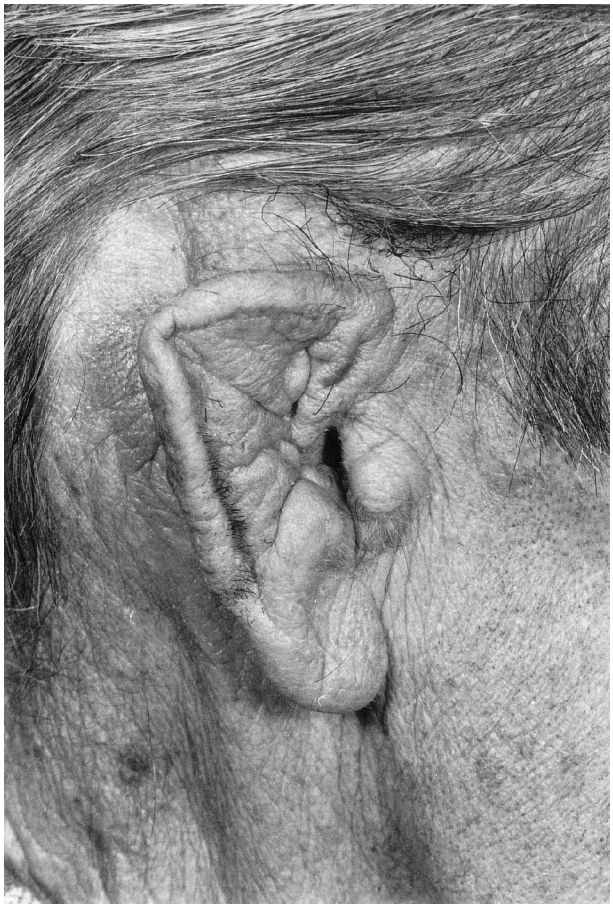

FIG. 1
Photograph of pinna showing the classic deformity seen in relapsing polychondritis.

no history of stridor. Fibre-optic nasendoscopy excluded any other focal pathology. In December 1998 the patient died following infective exacerbation of this COPD.

Discussion

Myelodysplastic syndomes (MDS) are clonal disorders of haematopoiesis characterized by peripheral cytopenias and dysplastic bone marrow, which is usually hypercellular.

Relapsing polychondritis is a rare disease characterised by inflammation and degeneration of the cartilage of the ears, eyes, nose, airways and joints. Clinical features include external ear chondritis, saddle nose deformity, subglottic strictures, arthritis, sensorineural hearing loss and otitis media. Relapsing polychondritis may at times represent a paraneoplastic phenomenon of MDS. MDS can precede relapsing polychondritis, can occur simultaneously or can develop during the evolution of relapsing polychondritis.[1,2]

McAdam[3] described criteria upon which the diagnosis of relapsing polychondritis can be made, in which patients must exhibit three of the following six features: bilateral auricular chondritis, non-erosive sero-negative inflammatory polyarthritis, nasal chondritis, ocular inflammation, respiratory tract chondritis and audiovestibular damage. This was later modified by Damiani[4] who stated that for diagnosis of relapsing polychondritis patients must have three clinical findings without histological confirmation, or chondritis of two anatomical sites that responds to steroids or dapsone. Histological examination of affected cartilage shows perichondral granulation tissue and inflammatory cell infiltration. The need for biopsy has been questioned because of the risk of infective perichondritis.[5]

CT and MRI have been advocated to aid the diagnosis of relapsing polychondritis.[6] Imaging is particularly useful in the diagnosis of laryngotracheobronchial involvement.[7] This may present with stridor, however, this was not a feature in our patient. CT is the first line of investigation. Typical findings are lumen narrowing, wall thickening and collapse of the supporting cartilaginous structures. Laryngeal cartilages can be partially calcified. It is felt that MRI should be used to complement CT, because of its long examination time and low anatomical resolution which is caused by image degradation secondary to respiratory motion.[7]

The cause of relapsing polychondritis is unknown. An autoimmune origin has been proposed and antibodies to collagen types II, IX and XI have been demonstrated.[8,9] Those demonstrated against type II collagen were mainly of the IgG subclass.[8] HLA tissue typing has shown a significantly increased frequency of antigen DR4.[10]

Thirty per cent of relapsing polychondritis cases have been found in association with other diseases including systemic vasculitis, systemic lupus erythematosus, rheumatoid arthritis, pulmonary fibrosis, polymyalgia rheumatica, ankylosing spondilitis, primary biliary cirrhosis and Hashimoto's disease. Relapsing polychondritis associated with MDS seems to be rare. In one study of 112 patients with relapsing polychondritis, only six had myelodysplastic or myeloproliferative syndromes.[2] However, in another bone marrow study of relapsing polychondritis, patients' evidence of myelodysplasia was quite frequent.[10]

From review of the literature, only one case of relapsing polychondritis with monoclonal gammopathy could be found[10] although MDS with monoclonal gammopathy is well documented.

Our case highlights the apparent existence of an association between relapsing polychondritis and MDS, which has not previously been reported in the otolaryngology literature. The possibility of a paraneoplastic phenomenon or the co-existence of systemic disease should be considered when patients are referred for primary assessment. This case illustrates the three features of myelodysplastic syndrome: monoclonal gammopathy and relapsing polychondritis in the same patient, which to our knowledge has not been described previously. The monocytosis had been present for at least 14 years and appeared to be increasing at the time of diagnosis of relapsing polychondritis but there is no convincing evidence that relapsing polychondritis is related to the magnitude or duration of monocytosis.

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