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Pathology in Focus

Mucous membrane plasmacytosis of the upper aerodigestive tract. A case report with effective treatment

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Abstract
We present a case of plasmacytosis of the mucous membrane of the upper aerodigestive tract. This is a rare benign condition characterized by plasma cell infiltration of the mucosa, with only nine cases described previously (Ferreiro *et al.*, 1994). The lesions, which have a cobblestone appearance, cause throat discomfort, dysphonia and mild dyspnoea. All the cases described previously failed to respond to antibiotics, systemic steroids, or to surgical resection. The present case has however responded favourably to intensive and prolonged treatment with beclamethasone oral spray and Corsodyl mouthwashes.

Key words: Plasmacytosis; Mouth; Pharynx; Larynx

Case report

A 69-year-old male former smoker, presented in July 1994 with a four-week history of left-sided throat discomfort, worse on swallowing, and a croaky voice.

Mirror examination of the throat revealed a cobblestone appearance of the left fauceal pillar and the laryngeal surface of the epiglottis. The left arytenoid appeared ulcerated with streaks of mucus. Two weeks later, on admission for biopsy and assessment under anaesthesia, the disease had progressed to involve the soft and hard palate (Figure 1), anterior third of the right vocal fold, middle third of the left vocal fold, left arytenoid, aryepiglottic fold and left posterior pharyngeal wall. Upper oesophagoscopy was normal. A chest X-ray and barium swallow did not reveal any abnormality. All blood tests including anti-neutrophil cytoplasmic antibodies (ANCA) were normal apart from a raised ESR of 60 mm/hr and an elevated anti-nuclear factor at 1.40 (IgM). The lesions were biopsied on two occasions and material sent for culture and histology. Cultures were negative for bacteria, AAFB and fungi. Both sets of biopsies showed similar histological appearances. The squamous epithelium showed striking hyperplasia and infiltration by acute and chronic inflammatory cells. There was a dense inflammatory infiltrate in the corium composed of plasma cells, together with some lymphocytes and macrophages (Figure 2). Stains for fungi and bacteria (in particular rhinoscleroma) and protozoa were negative and repeated immunohistochemistry for light chains showed the plasma cells to be polyclonal. The immunohistochemistry was confirmed by molecular biological studies, which showed no clonal rearrangement of the Jh genes.

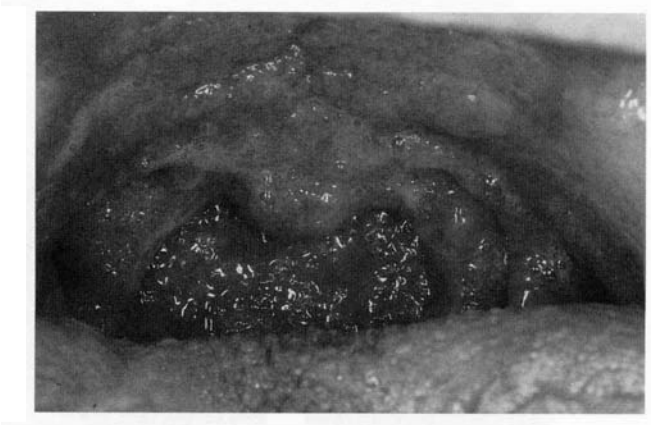

FIG. 1
Cobblestone appearance of the hard and soft palate and the posterior pharyngeal wall.

Whilst the final pathological diagnosis was awaited the patient was allowed home, but was readmitted three months later with worsening of his dysphagia, hoarseness, and haemoptysis. Fibreoptic examination revealed the lesions in his larynx and hypopharynx to be more florid than previously. He was commenced on Corsodyl mouthwashes and a beclamethasone spray, two puffs six hourly, applied locally to the oral cavity and pharynx rather than inhaled. There was immediate relief in his symptoms. Since then he has continued to improve on a reduced dose of beclamethasone two puffs twice daily and twenty months after presentation now shows no lesions in his oropharynx, larynx nor hypopharynx.

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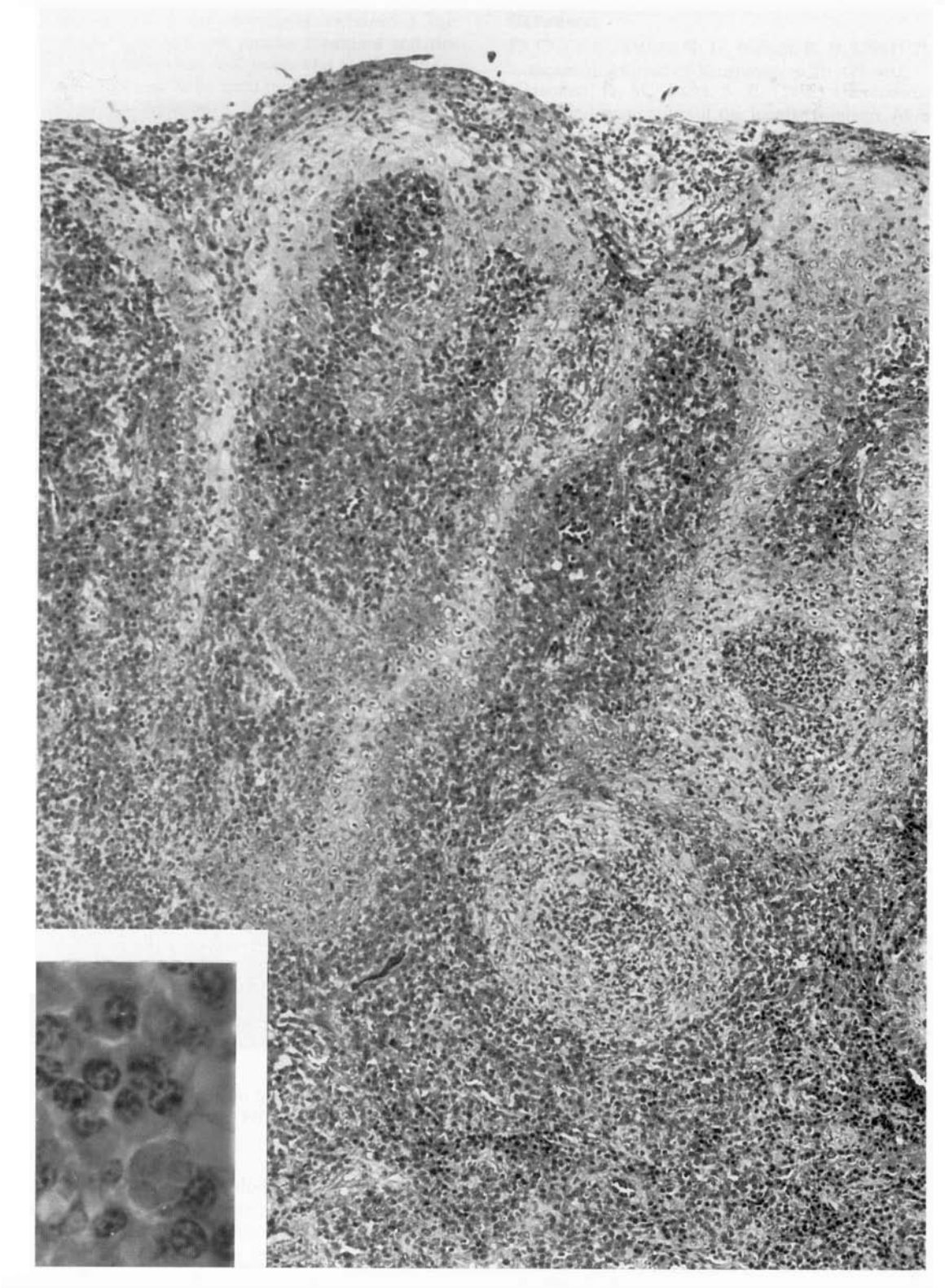

FIG. 2

Striking epithelial hyperplasia with a heavy inflammatory infiltrate in the epithelium and connective tissue. (H & E; × 100). Inset (× 640) shows characteristic plasma cell morphology, including Russell bodies.

Discussion

Mucous membrane plasmacytosis is an unusual non-neoplastic condition of plasma cell proliferation, the aetiology, pathogenesis and treatment of which is not known. The disease affects the upper aerodigestive tract mucosa with multiple cobblestone lesions with normal mucosa in between. Occasionally the lesions may be ulcerative.

Clinically, the condition is very rare and the diagnosis is made by exclusion of similar conditons like extramedullary plasmacytoma, sarcoid, rhinoscleroma, cicatricial pemphigoid, lichen planus and fungal conditions which can affect

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the upper aerodigestive tract. Extramedullary plasmacytoma commonly occurs in the head and neck, mainly in older men and shows preferential involvement of the supraglottic larynx (Horny and Kaiserling, 1995). The lesions are mainly polypoidal and non-ulcerative. There is minimal atypia of plasma cells on histology, but the plasma cells are monoclonal. Sarcoid of the larynx affects mainly the supraglottic and subglottic regions, which show pale diffuse areas of mucosal thickening and a granulomatous infiltrate on histology.

Benign mucous membrane pemphigoid (cicatricial pemphigoid) in common with the mucous membrane plasmacytosis, affects the elderly (Foster and Nally, 1977; Hardy *et al.*, 1971). The condition can affect the nasal mucosa, nasopharynx, larynx and hypopharynx. However, this is a condition affecting mainly women with ulceration which heals with scarring. In contrast, the lesions in plasmacytosis are mainly raised and cobblestone.

Laryngeal scleroma may also affect the nose, larynx and trachea. The disease is caused by *Klebsiella rhinoscleromatis* and affects a younger age group of either sex. Scleroma is more commonly seen in parts of Central and South America, Russia, Central Europe, parts of Africa, Egypt and Pakistan. Histologically, the disease is characterized by a plasma cell infiltrate in the submucosa with foamy macrophages (Mikulicz cells). The plasma cells may contain Russell bodies (inspissated immunoglobin). The Mikulicz cells stain strongly with the periodic acid-Schiff reaction (PAS). Neither convincing Mikulicz cells nor Russell bodies were present in this case.

Lichen planus is another inflammatory condition affecting the skin and mucous membrane. In the mouth the lesions appear as white patches, typically with a lace-like pattern. The lesions may cause pain. Microscopically, in the skin there is striking hyperkeratosis in the upper dermis and lower epidermis showing a superficial, band-like infiltration with lymphocytes. Mucosal lesions show the band-like inflammatory infiltrate but the hyperkeratosis may not be so apparent. Erosive changes may also be seen.

The treatment of mucous membrane plasmacytosis is not established. A variety of treatments including antibiotics, systemic steroids and surgical or carbon dioxide laser resection have been tried to shrink the lesions. In two cases reported (Ferreiro *et al.*, 1994) the disease had progressed, causing sufficient airway obstruction to require a tracheostomy. Although none of the previously reported cases have shown any signs of disease regression, the long-term survival is good with the longest survivor having 16 years with the disease (Ferreiro *et al.*, 1994).

Conclusion

Mucous membrane plasmacytosis of the aerodigestive tract is a non-neoplastic condition which can have disabling effects on the airway. A variety of medical and surgical treatments have been tried but none has led to improvement of the condition. This is the first case where treatment has been shown to be associated with regression of the condition.

Acknowledgement

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