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Laryngeal malakoplakia

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Abstract
We report a case of malakoplakia of the epiglottis in a 45-year-old female patient. Only three cases of laryngeal malakoplakia have been reported in the world literature, one of which was associated with tracheal malakoplakia. To our knowledge this is the first reported case of isolated malakoplakia of the epiglottis. Malakoplakia, a rare granulomatous lesion, has been seen more frequently in the urinary tract. Other organs like the genito-urinary tract, testis, epididymis, lymph nodes, middle ear, nasopharynx, tonsil and retroperitoneal tissue have been also involved though less frequently.

Key words: Granuloma, respiratory tract; Epiglottis

Introduction

Malakoplakia is a rare granulomatous lesion which may mimic a neoplasm. The diagnosis is made only on histopathological examination (Benizri *et al.*, 1993). Of all cases of malakoplakia described, 75 per cent have been reported in the urinary tract (Stanton and Maxted, 1981). The aetiology of the condition is not known. This disease is characterized by tumour-like lesions which on histological examination show an accumulation of macrophages the von-Hansemann's cells which contain calcified cytoplasmic inclusions known as the Michaelis-Gutmann bodies (Michaelis and Gutmann, 1902; Lou and Teplitz, 1974). The macrophages also contain undigested coliform bacilli and although the pathological origin of malakoplakia is still controversial, the consensus opinion is that malakoplakia represents a chronic inflammatory reaction to an infectious agent particularly *Escherichia coli* (Long and Althausen, 1989). Malakoplakia can have significant morbidity and mortality if vital organs are involved.

Case report

A 45-year-old female patient, a non-smoker presented with a history of hoarseness of voice, painful throat and difficulty on swallowing for six months. The patient had no other systemic problems or symptoms or signs of decreased immunity. Laboratory studies revealed a haemoglobin of 10.8 gm%. The total white blood cell count was elevated to 22×10^9/l with 90 per cent neutrophils. The ESR was also raised to 50 mm/1st hour. The blood chemistry was unremarkable. Test for the human immunodeficiency virus (HIV) was negative. Urine examination was negative for sugar albumin and microscopy revealed no abnormality. The chest X-ray was normal.

On ENT examination a proliferative growth of the epiglottis was noted. Clinical examination of the chest and abdomen revealed no abnormality. Direct laryngoscopic examination showed a nodular lesion on the laryngeal surface of the epiglottis. The vocal folds, vestibular folds, pyriform fossae and subglottis were normal.

Bronchoscopic examination revealed no abnormality of the bronchial tree. The lesion was excised and sent for histopathological examination, with suspicion of a malignancy. The patient was given antibiotics post-operatively.

Histopathological examination

The biopsy consisted of a polypoid structure lined by stratified squamous epithelium. The subepithelium was infiltrated by mixed inflammatory cells including polymorphonuclear leukocytes, lymphocytes, plasma cells and sheets of histiocytes (Figure 1).

The histiocytes had abundant eosinophilic, coarsely granular and occasionally vacuolated cytoplasm. Occasional giant cells were also present. The histiocytes contained one or more round weakly basophilic, centrally laminated, intracytoplasmic inclusions measuring up to 10 mm in size, namely Michaelis-Gutmann bodies (Figure 2). These bodies stained positively with periodic acid-Schiff

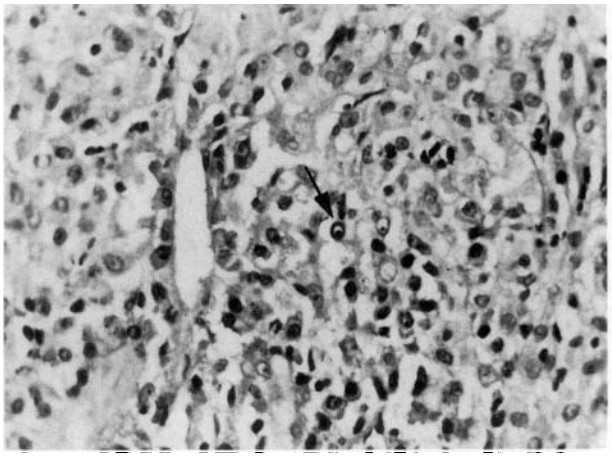

FIG. 1
Histology showing mixed inflammatory cells (H & E; × 550).

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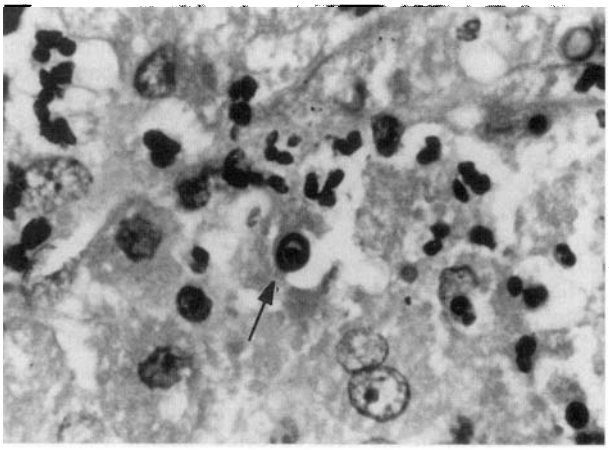

FIG. 2
Histology showing Michaelis-Gutmann bodies (H & E; × 1100).

(PAS), VonKossa (for calcium) and Pearl's reaction (for iron).

Discussion

Laryngeal malakoplakia has been described previously in only three patients. One case reported by Mollo *et al.* (1994) was associated with tracheal malakoplakia. The present report is the first case of isolated malakoplakia of the epiglottis. In our patient due to her age and duration of the symptoms, malignancy was suspected. Malakoplakia was diagnosed only on histopathological investigation.

The patient had a raised total leucocyte count and neutrophilia suggesting an acute infectious aetiology. In patients associated with urinary tract malakoplakia Gram negative bacilli such as *Escherichia coli* and *Klebsiella* have been demonstrated and these are considered to reach other sites through the blood (Saleem *et al.*, 1993). Our patient did have fever before the symptoms appeared, but no urine analysis was done at that time and subsequent urine analysis was normal. Malakoplakia has been described in individuals with decreased immune activity, malignancy and other systemic disorders (Streem, 1984; Witherington *et al.*, 1984). Our patient had none of these conditions.

Various authors have reported malakoplakia in which the phagocytic activity of the peripheral leukocytes was normal, but the intracellular cyclic guanosine monophosphate level was reduced, associated with a decreased release of β-glucoronidase and weaker bactericidal activity. Therefore it was suggested that the basic defect in malakoplakia might be the functional abnormality of the leukocyte (Abdou *et al.*, 1977).

The diagnosis of malakoplakia is based on histological examination characterized by an inflammatory infiltrate consisting mainly of eosinophilic histiocytes and PAS positive intracytoplasmic inclusions (Michaelis and Gutmann, 1902). These inclusions are phagolysosomes enclosing improperly digested bacteria. In the early phase there is prevalence of plasma cells and in the late stage there is a preponderance of fibroblasts and collagen fibres with rare Michaelis-Gutmann bodies (Flint and Mured, 1984).

It is interesting to note that our patient had no predisposing factors such as smoking cigarettes or any other associated condition.

The management of malakoplakia localized to the lower urinary tract is conservative (Cozar *et al.*, 1993). In accessible regions surgery shortens the duration of medical treatment. Disseminated malakoplakia has been treated successfully by Van Furth *et al.* (1992) with ciprofloxacin (500 mg b.i.d.), a fluoroquinolone. Several months seem to be necessary for cure. Cholinergic agonists, like bethanecol which are supposed to enhance macrophage bactericidal activity through raising the cytoplasmic cyclic guanosine monophosphate level have been employed (Oliver, 1976). In our patient during direct laryngoscopy, the tumour was excised and antibiotic treatment was started after the histological report was available. The patient was given ciprofloxacin for six weeks after which she has been followed-up for two years with no further development of any lesion and urinanalysis showed no abnormality.

References

Abdou, N. I., Pombejara, C. N., Sagawa, A. (1977) Malakoplakia: evidence for monocyte lysosomal abnormality correctable by cholinergic agonist in vitro and in vivo. *New England Journal of Medicine* **297:** 1413–1419.

Benizri, E., Hofman, P., Chevallier, D., Fabiani, P., Suhler, A., Peyrottes, A., de Graeve, B., Marie, J. F., Volpe, P., Amiel, J. (1993) Urinary malacoplakia. Anatomoclinical analysis of three cases with long-term follow-up. *Journal d'Urologie* **99 (1):** 38–43.

Cozar Olmo, J. M., Carcamo, P., Gaston 'de Iriarte', E., Jimenez, F., Martinez-Pineiro, L., Martinez-Pineiro, J. A. (1993) Genitourinary malakoplakia. *British Journal of Urology* **72(1):** 6–12.

Flint, A., Mured, T. M. (1984) Malakoplakia and malakoplakia-like lesions of the upper gastrointestinal tract. *Ultrastructural Pathology* **7:** 167.

Long, J. P., Althausen, A. F. (1989) Malacoplakia: a 25-year experience with a review of the literature. *Journal of Urology* **141:** 1328–1331.

Lou, T. Y., Teplitz, C. (1974) Malakoplakia: pathogenesis and ultrastructural morphogenesis. *Human Pathology* **5:** 191–207.

Michaelis, L., Gutmann, C. (1902) Uber Einschlusse in Blasentumoren. *Journal of Clinical Medicine* **47:** 208–215.

Mollo, J., Groussard, O., Baldeyrou, P., Molas, G., Fournier, M., Pariente, R. (1994) Tracheal malacoplakia. *Chest* **105:** 608–610.

Oliver, J. M. (1976) Impaired microtubule function correctable by cyclic GMP and cholingeric agonist in the Chedial Higashi syndrome. *American Journal of Pathology* **85:** 395.

Saleem, M. A., Milford, D. V., Raafat, F., White, R. H. R. (1993) Renal parenchymal malakoplakia: a case report and review of the literature. *Pediatric Nephrology* **7:** 256–258.

Stanton, M. J., Maxted, W. (1981) Malakoplakia: a study of the literature and current concepts of pathogenesis, diagnosis and treatment. *Journal of Urology* **125:** 139–146.

Streem, B. S. B. (1984) Genitourinary malacoplakia in renal transplant recipients: pathogenic, prognostic and therapeutic considerations. *Journal of Urology* **132:** 10–12.

Van Furth, R., van't Wout, J. W., Wertheimer, P. A., Zwartendijk, J. (1992) Ciprofloxacin for treatment of malakoplakia. *Lancet* **339:** 148–149.

Witherington, R., Branan, W. J., Wray, B. B., Best, G. K. (1984) Malakoplakia associated with vesicoureteral reflux and selective immunoglobulin A deficiency. *Journal of Urology* **132:** 975–977.

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