

INTERSTITIAL KERATITIS AND DEAFNESS IN A PATIENT WITH CUTANEOUS SARCOIDOSIS

DE SMEDT S. *, AYLIFFE W. *

SUMMARY

We report a case of interstitial keratitis and progressive hearing loss in a young female patient with biopsy proven cutaneous sarcoidosis. This rare sequence of ophthalmological and auditory signs in sarcoidosis mimicks Cogan's syndrome.

SAMENVATTING

We stellen een casus voor van interstitiële keratitis en progressief gehoorverlies bij een jonge vrouw met anatomopathologisch bewezen cutane sarcoidosis. Deze zeldzame combinatie van oftalmologische en auditieve bevindingen in sarcoidosis bootst Cogan syndroom na.

RÉSUMÉ

Nous présentons un cas de kératite interstitielle et surdité progressive chez une patiente présentant une sarcoidose cutanée confirmée anatomopathologiquement. Cette combinaison rare de signes ophtalmologiques et auditifs dans le cadre de la sarcoidose, mime le syndrome de Cogan.

KEY-WORDS

Sarcoidosis, interstitial keratitis, hearing loss, Cogan's syndrome

MOTS-CLÉS

Sarcoidose, kératite interstitielle, surdité progressive, syndrome de Cogan

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* Croydon Eye Unit, University Hospital, Croydon CR7 7YE, United Kingdom

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INTRODUCTION

Sarcoidosis is a chronic, multisystem, granulomatous disease. Interstitial keratitis is an uncommon finding. Also only a few cases of hearing loss associated with sarcoidosis have been documented. This paper describes an unusual association of biopsy proven cutaneous sarcoidosis, hearing loss and interstitial keratitis in a young woman, mimicking Cogan's syndrome.

CASE

A 37-year-old female with a marked facial rash presented to the eye clinic with blepharitis and marginal corneal scarring. She was diagnosed as having acne rosacea and treated with oxytetracycline 250 mg and prednisolone 1% drops twice a day. Three months later, a conjunctival nodule and episcleritis appeared in the left eye and both resolved on the same treatment.

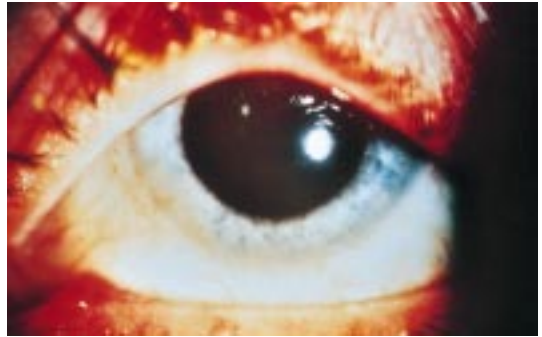


Fig 1: Corneal photograph showing a dusky pink lesion of the peripheral cornea (salmon patch) corresponding to active interstitial keratitis at 7 o'clock.

Six months after initial presentation she developed a painful inflamed left eye. Visual acuity was 6/6 right, 6/9 left. A deep stromal vascularisation of the peripheral cornea at 7 o'clock was seen (figure 1).

There was no history of fever, fatigue, weight loss, pulmonary nor meningeal symptoms. Cutaneous sarcoidosis had been diagnosed two

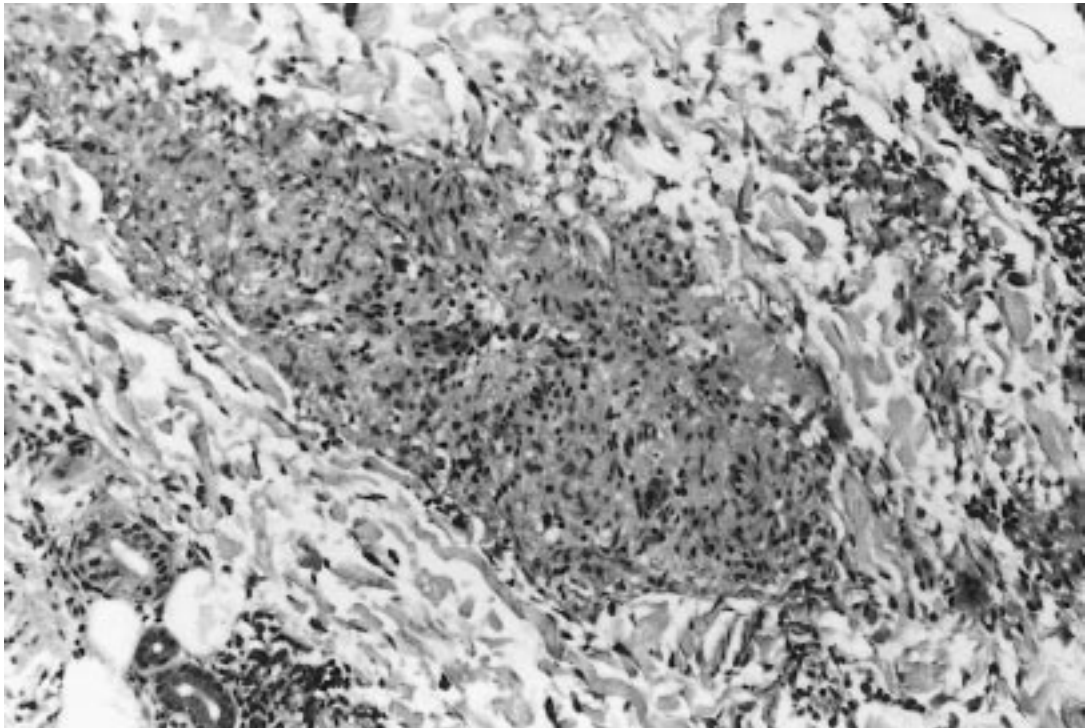


Fig 2: This photomicrograph shows a skin biopsy with non-caseating epithelioid granulomas in the dermis. (haematoxylin and eosin, original magnification X630).

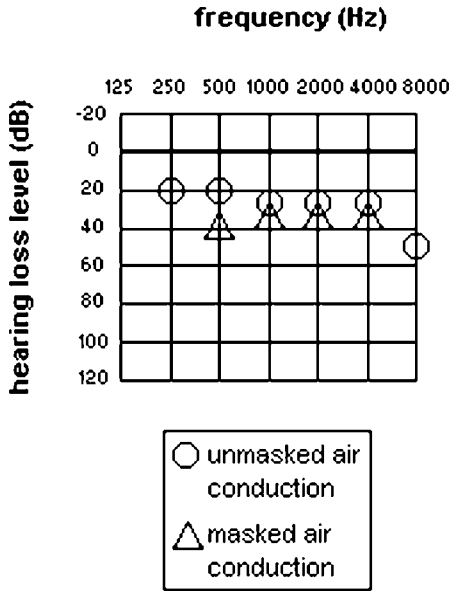


fig 3a: right ear audiogram

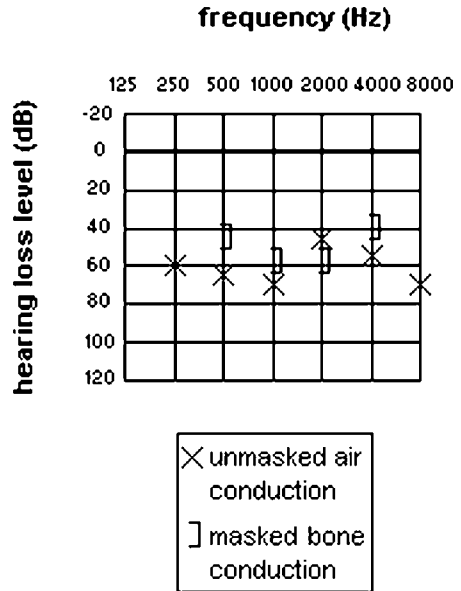


fig 3b: left ear audiogram

Fig 3: Audiogram showing a mild high frequency loss on the right (fig 3A), and a moderate to severe sensorineural hearing loss on the left (fig 3B). (normal range 0-20dB).

years previously on the basis of a skin biopsy showing typical non-caseating epithelioid granulomas (figure 2). Gradual hearing impairment had been developed since the last two years. There was no history of vertigo or tinnitus.

Investigations including a chest X-ray, full blood count, erythrocyte sedimentation rate, C-reactive protein, serum angiotensin converting enzyme (sACE), rheumatoid factor, anti-nuclear antibodies, anti-neutrophilic cytoplasmic antibody, syphilis serology and complement levels were normal. Audiometry revealed a moderate to severe sensorineural hearing loss on the left and a mild high frequency loss on the right (figure 3). A MRI of her head was normal.

The interstitial keratitis responded to topical prednisolone drops six times a day. Systemic steroids were not used because recovery of hearing was unlikely.

DISCUSSION

Sarcoid is an ideopathic, chronic, multisystem, granulomatous disease that predominantly af-

fects the lungs, thoracic lymph nodes, skin, and eyes. Some of the more commonly cited ocular findings include: lacrimal gland swelling, orbital granulomas, conjunctival granulomas, anterior uveitis and its sequelae, vitritis, periphlebitis, chorioretinitis, optic disc edema, and optic nerve involvement (6). Corneal involvement is extremely rare (4), but interstitial keratitis has been reported (6). Only a few cases of hearing loss associated with sarcoidosis have been documented (2,5,8,11). Since there are no definitive diagnostic tests, the presence of non-caseating granulomas on tissue biopsy together with compatible clinical features is usually considered as a proof of diagnosis of sarcoidosis (4,10). In this case report, a diagnosis of cutaneous sarcoidosis was histopathologically made two years prior to ocular presentation. ACE-levels were normal and although it might reflect the disease activity, normal levels do not exclude the diagnosis of sarcoidosis (4,10).

Interstitial keratitis refers to nonsuppurative infiltration and vascularisation of the deep corneal stroma and is associated with conditions such as syphilis, systemic lupus erythemato-

sus and viral infections such as mumps, rubella and herpes. The marginal form, a so called salmon patch, is unlikely to progress to the more, classic interstitial keratitis after topical steroid use (7).

Laboratory tests and a chest X-ray ruled out other conditions. Only subepithelial marginal corneal infiltration has been reported in acnea rosacea (3).

Conjunctival nodules and episcleritis as reported in this case report, also occur in sarcoidosis (10).

The interstitial keratitis in combination with sensory neural hearing loss is suggestive of Cogan's syndrome. However, the classic audio-vestibular dysfunction is acute in onset, characterized by Menière-like attacks with progressive hearing loss (12). Nevertheless cases with hearing loss only have been reported (7). Any intracranial cause for the hearing loss, such as acoustic neuroma, has been excluded by the MRI Scan.

The vestibulocochlear nerve is the fourth most frequently affected cranial nerve in sarcoidosis (1,4,9,12). There are no specific diagnostic tests for neurosarcoidosis. ACE-levels do not appear to reflect the activity of neurological disease and cerebrospinal fluid findings are variable and non-specific (1). The diagnosis of neurosarcoidosis demands a compatible clinical picture of a multisystem disease and histological confirmation of sarcoidosis tissue. The loss of hearing is typically sensorineural and sudden in onset or slowly progressive (12). The audiovestibular dysfunction reported in sarcoidosis becomes irreversible resulting from ischaemia due to vasculitis (9). This could be the mechanism by which sarcoidosis caused a 'Cogan's syndrome- picture' in this patient. Furthermore, Cogan's syndrome has occurred in association with acute sarcoidosis (7). However, this is the first case of cutaneous sarcoidosis, mimicking Cogan's syndrome.

CONCLUSION

This case demonstrates the rare association of cutaneous sarcoidosis with interstitial keratitis and deafness, mimicking Cogan's syndrome. The tissue diagnosis of sarcoidosis was obtained on skin biopsies.

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Correspondence
W. Ayliffe, Croydon Eye Unit, University Hospital,
Croydon CR7 7YE, U.K.