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Clinical Record

Sarcoidosis of the external ear – literature review and report of a case

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Abstract
External ear manifestations of sarcoidosis are rare. We review six cases in the literature and also report a case. The otolaryngologist plays an important role in making the diagnosis because of the ease of biopsy in all cases of sarcoidosis of the external ear.

Key words: Sarcoidosis; Ear, External; Ear Piercing

Introduction

Sarcoidosis was described by the World Congress in Kyoto in 1991 as 'A multisystemic disorder of unknown cause. It commonly affects young and middle-aged adults and frequently presents with bilateral hilar lymphadenopathy, pulmonary infiltration, ocular and skin lesions. Other organs may be involved. The diagnosis is established when clinicoradiological findings are supported by histological evidence of non-caseating epitheloid cell granulomas. Granulomas of known cause and local sarcoid reactions must be excluded.'[1]

Otolaryngological manifestations of sarcoidosis occur in 10–15 per cent of patients and only rarely are the presenting disorder.[2] Cervical adenopathy is the most common head/neck manifestation of sarcoidosis and was present in 48 per cent in one series.[3] Parotid gland involvement occurs in 6 per cent of patients and is usually bilateral.[4] Sinonasal manifestations occur in 1–4 per cent of patients[5,6] while neurosarcoidosis is found in about 5 per cent with the most commonly affected cranial nerve being the facial nerve.[7] The larynx is affected in 0.5–8.3 per cent of patients[8,9] and tonsillar involvement is found in 2.4 per cent.[10]

The ear is, however, rather an uncommon site although there have been anecdotal reports of middle-ear involvement[11] and acoustic nerve with sensorineural deafness.[12] Despite the fact that skin involvement occurs in between 20–35 per cent of patient with systemic disease,[13] cutaneous sarcoidosis of the external ear is rarely reported. The purpose of this paper is to collect reported cases and present the case of an additional patient.

Historical review

Historically, it is of interest to note that Hutchinson is credited with the first description of sarcoidosis in 1875.[14] He named it after one of his patients, who had unique skin findings, as 'Mortimer's Malady'. In 1889, Besnier in his paper described sarcoid lesions of both ears and nose.[15] He showed similarity of the lesions to lupus, pointing to the reddish-purple nodular tumours. However, in 1899 Cae Sar Boeck first coined the term sarcoid to describe one of the skin lesions of sarcoidosis because of its histological resemblance to sarcoma.[14] In 1909, Danish ophthalmologist Heerfordt described the triad of uveitis, parotid enlargement and cranial nerve paresis.[16] The pathological findings were described in 1916 by Shaumann.[17]

Methods

A systematic review of the medical literature was conducted. Articles were identified using MEDLINE (1966–2004). The search strategy used Medical Subject Heading (MeSH) terms 'Sarcoidosis and Ear', 'ENT and Sarcoidosis' and 'Cutaneous Manifestations of Sarcoidosis'. The titles and abstracts of articles identified were examined and independently reviewed. References from retrieved articles were reviewed to identify additional pertinent articles. Articles with reports of cutaneous sarcoid reactions secondary to foreign material, and development of sarcoid lesions in scars and tattoos were excluded from review.

Results

The literature search identified four articles and two others were retrieved from reference articles. Poe[18] reported one of the earliest cases in England. It was that of a 38-year-old woman with widespread sarcoid disease. She presented with multiple, small, non-tender nodules. These were located over the entire external ear including the lobule, they were numerous and about the size of a pinhead.

Boone and Coleman[19] made another report. This was a case of a man of 40 years old with systemic and multiorgan involvement. Several large nodules were present on both auricles and the resulting distortion was similar to the disfigurement seen in leprosy. Cavallaro[20] reported a case of a 70-year-old woman with a thickened and erythematous

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right ear lobe. Several physicians had treated it as eczema. As with other cases, a biopsy of the lesion confirmed sarcoidosis. In spite of not having systemic symptoms a chest X-ray showed widened mediastinum with hilar lymphadenopathy consistence with sarcoidosis.

Nova[21] described the case of a nine-year-old boy with a six month history of a slowly enlarging mass of his left superior helix. On examination of the lesion there was a raised nodular mass with superficial crusting with adherence to the underlying perichondrium. The boy had no systemic symptoms. Mann and Peachey[22] reported the case of a 14-year-old girl with multiple soft, reddish-brown coloured nodules on both ear lobes at the site of recent piercings. There was no systemic involvement. Biopsy confirmed sarcoidosis with a positive Kveim test. There was no foreign material seen in the biopsy.

Lang *et al.*[23] reported recently on a 38-year-old man with an acute exacerbation of swelling of the right pinna. There were associated systemic symptoms. They found inflamed and thickened right helical cartilage and pinna with overlying skin being erythematous and excoriated. Biopsy of the lesion and a chest X-ray were consistent with pulmonary sarcoidosis.

Report of a case

A general practitioner referred a 30-year-old man to us with an inflammatory lesion on the left lobule. This had been present for seven months and had been treated several times with antibiotics by the general practitioner with no response. The patient had his left ear pierced by a jeweller one month prior to noticing the inflammatory lesion at the site of recent piercing. There was no past history of tuberculosis or contact with persons with chronic cough. It is of note that the patient works in a factory that manufactures silicone-based medical prostheses.

On examination we found bilaterally non-tender moderately enlarged upper cervical lymphadenopathy with an enlarged left submandibular gland. Examination of the lesion showed a reddish plaque with underlying inflammatory exudates at the site of recent piercing (Figure 1). It was noted that he had other piercings, which have been present for some years and these were not involved. There did not appear to be any foreign body material in the lobule. Other ENT examinations showed bilaterally enlarged tonsils but no inflammation. Systemic examination was normal.

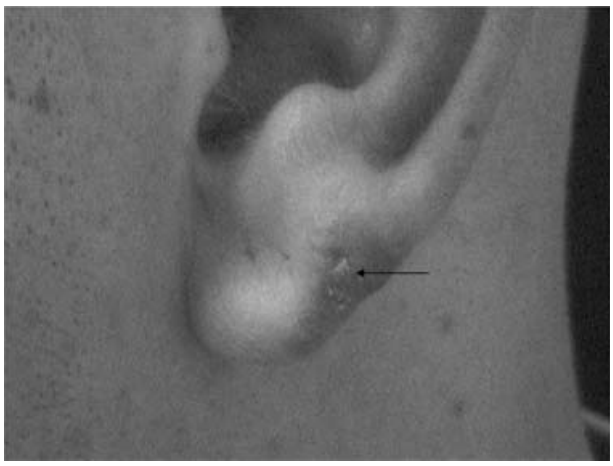
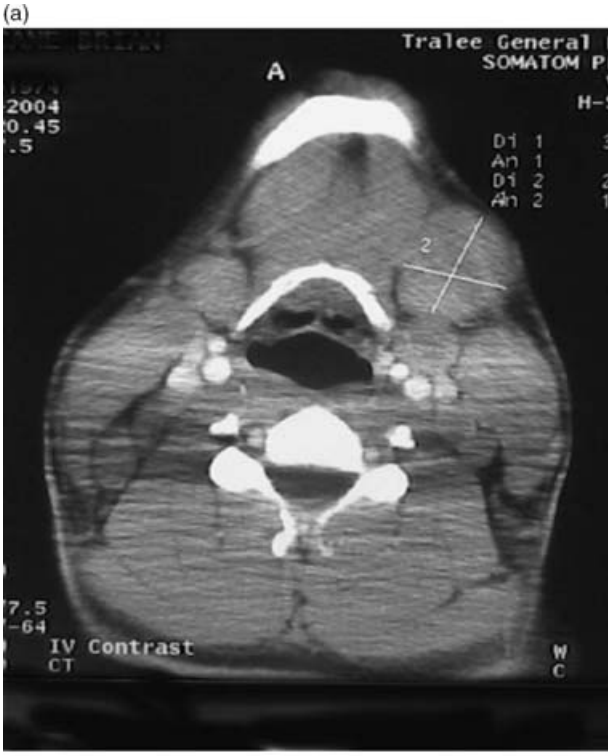


FIG. 1

The arrow points to the area of reddish plaque which has the underlying inflammatory exudates at the site of recent piercing (magnification ×20).

Routine blood tests revealed mildly elevated serum angiotensin converting enzyme (ACE), other parameters were normal. Autoimmune antibody testing was negative. Testing for fungi and mycobacteria were negative. Also viral cultures were negative. However, the serum was positive for mycoplasma pneumonia. We noted that the chest X-ray was reported as normal. As the lesions did not appear to be settling we decided to arrange a computed tomography (CT) scan of the neck and thorax and biopsy of the lesion. The CT scan of the neck confirmed cervical lymphadenopathy and the CT scan of the thorax showed widespread pulmonary infiltrates with hilar and mediastinal lymphadenopathy consistent with pulmonary sarcoidosis. (Figure 2a and 2b)

(a)



(b)

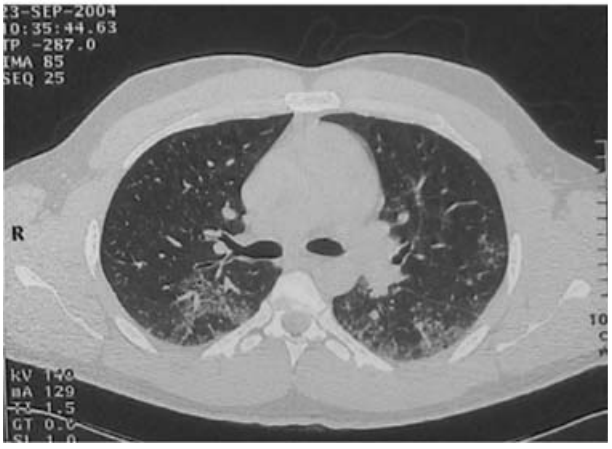


FIG. 2

(a) CT of the neck showing cervical lymphadenopathy (magnification ×20). (b) CT of the thorax showing widespread pulmonary infiltrates with hilar and mediastinal lymphadenopathy (magnification × 20).

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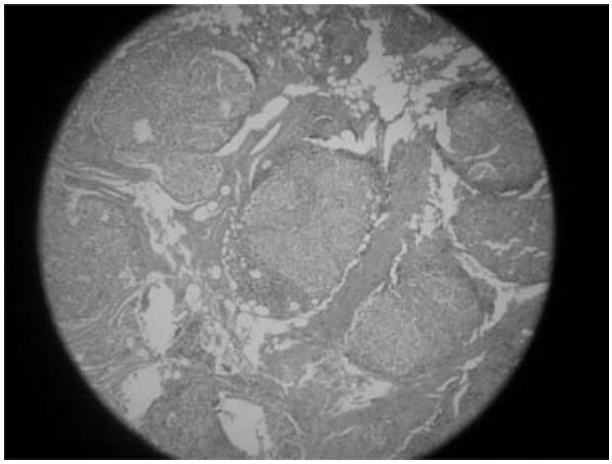

FIG. 3
Histology of the skin showing non-caseating epitheloid cell granulomata (H&E; ×200).

Histology of the biopsy of the lesion identified non-caseating epitheloid cell dermal granulomas consisting of macrophages, multinucleate giant cells and lymphocytes, findings consistent with sarcoidosis (Figure 3). No foreign body materials were seen in the skin biopsy. Special stains for acid-fast bacilli and fungi were negative.

- **Sarcoidosis of the external ear is rarely reported. Its manifestations vary and mimic most common diseases of this area. A high index of suspicion is important and otolarygologists should include this entity in the differential diagnosis of lesions on the external ear**
- **Biopsy of specimens should be obtained from the most readily accessible organ with the least invasive method. Thus the otolaryngologist is critical to diagnosis because of the ease of biopsy in this area. The Kveim test is no longer used to make the diagnosis of sarcoidosis**
- **Corticosteroids remain the mainstay of treatment, though a wide range of alternative systemic drugs have been employed**

The patient was referred to a respiratory physician for further management. He was placed on oral corticosteroids. A review of the patient after one month showed regression of the skin lesion and neck lymphadenopathy. In this case we did not perform the Kveim–Siltzbach skin test because it is no longer approved for general use.[24] Also having made a clinicoradiographic and histological diagnosis of sarcoidosis, a bronchoalveolar lavage or transbronchial biopsy were not necessary to confirm diagnosis as these are invasive procedures.

Summary

Sarcoidosis of the external ear is rarely reported. Its manifestations vary and mimic most common diseases of this area, so a high index of suspicion is important and the otolaryngologist should include this entity in their differential diagnosis of lesions on the ear. Biopsy of specimens should be obtained from the most readily accessible organ with the least invasive method.[25] Thus the otolaryngologist is critical to diagnosis because of the ease of biopsy in this area. The Kveim test is no longer used to make the diagnosis of sarcoidosis.

Corticosteroids remain the mainstay of treatment, although a wide range of alternative systemic drugs have been employed. The reports of their use are often anecdotal. Such agents include hydrochloroquine, thalidomide and allopurinol, and immunosuppressives like methotrexate[24–28] are in use for treatment and these may be successful alternatives to the use of steroids. Although the literature review showed that ear piercing could predispose the patient to a sarcoid lesion, we thought our observed case might have been a case of Koebner phenomenon occurring in systemic sarcoidosis. We found no report of this association in the literature. Also there is evidence in the literature that sarcoid lesions can occur in surgical scars, tattoos, and the site of venepuncture and vaccination.[29–32] Scar sarcoidosis may either occur in isolation or parallel systemic disease.[33] It has been shown that sarcoidosis can occur in the sites of ear piercing, therefore it is advisable that patients with sarcoidosis should avoid piercing their ears.

References

1 Yamamoto M, Sharma OP, Hosoda Y. Special report: the 1991 descriptive definition of sarcoidosis. *Sarcoidosis* 1992;**9**(Suppl 1):33–4
2 Shah UK, White JA, Gooey JE, Hybels RL. Otolaryngologic manifestations of sarcoidosis: presentation and diagnosis. *Laryngoscope* 1997;**107**:67–75
3 Dash GI, Kimmelman CP. Head and Neck manifestations of Sarcoidosis. *Laryngoscope* 1988;**98**:50–3
4 James DG, Sharma OP. Parotid gland sarcoidosis. *Sarcoidosis Vasc Diffuse Lung Dis* 2000;**17**:27–32
5 McCaffrey TV, McDonald TJ. Sarcoidosis of the nose and paranasal sinuses. *Laryngoscope* 1983;**93**:1281–4
6 Zeitlin JF, Tami TA, Baughman R, Winget D. Nasal and sinus manifestations of sarcoidosis. *Am J Rhinol* 2000;**14**: 157–61
7 Delaney P. Neurologic manifestations in sarcoidosis: review of the literature with a report of 23 cases. *Ann Intern Med* 1977;**87**:336–45
8 Gallivan GJ, Landis JN. Sarcoidosis of the larynx, preserving and restoring airway and professional voice. *J Voice* 1993;**7**:81–94
9 Noel HB III, McDonald TJ. Laryngeal sarcoidosis: a report of 113 patients. *Ann Otol Rhino Laryngol* 1980;**89**(1Pt1): 58–61
10 Maycock RL, Bertrand P, Morrison CE. Manifestations of sarcoidosis. *Am J Med* 1973;**35**:67–89
11 Tyndel FJ, Davidson GB, Birman H, Modzelewski ZA, Acker JJ. Sarcoidosis of the middle ear. *Chest* 1994;**5**: 1582–3
12 Colover J. Sarcoidosis with involvement of the nervous system. *Brain* 1948;**71**:451–75
13 Kadel FA, Moschella SL. Sarcoidosis: an updated review. *J Am Acad Dermatol* 1984;**11**:1–19
14 Hunter FT. Hutchinson-Boeck's disease (generalised sarcoidosis): Historical note and report of a case with apparent cure. *N Eng J Med* 1936;**214**:346
15 Besnier E. Lupus of the face. *Ann of Derm Syph* 1889;**10**: 333–36
16 Heerfordt CF. Uveoparotid fever [in German]. *Albrecht von Graef's Arch Ophthalmol* 1909;**70**:254
17 Shaumann JE. Study on pernio lupus and its reports with sarcoidosis and tuberculosis [in French]. *Ann Dermatol Syphil* 1916;**6**:357
18 Poe DL. Sarcoidosis of the external ear. *Ann Otolaryng* 1940;**49**:771–5
19 Boone JA, Coleman RR. Sarcoidosis. *Southern Med J* 1944;**37**:477–81
20 Cavallaro AG. Sarcoidosis of the external ear. *Arch Otolaryng* 1969;**90**:293–5
21 Nova S. Sarcoidosis of the ear. *Ear Nose Throat J* 1981;**60**: 387–8

https://doi.org/10.1017/S0022215106003902 Published online by Cambridge University Press

22 Mann RJ, Peachey RDG. Sarcoid tissue reaction – another complication of ear piercing. *Clin Exp Dermatol* 1983;**8**: 199–200
23 Lang EM, Zaruk JE, Colreavy MP, Kennedy S, Rowley H, Timon C. An unusual case of external ear inflammation caused by sarcoidosis. *Ear Nose Throat J* 2003;**82**:942–5
24 Zic JA, Horowitz DH, Arzubiaga C, King LE Jr. Treatment of cutaneous sarcoidosis with Chloroquine: review of the literature. *Arch Dermatol* 1991;**127**:1034–40
25 Newmann LS, Rose CS, Maier LA. Sarcoidosis. *N End J Med* 1997;**24**:1224–34
26 Carlesimo M, Giustini S, Rossi A, Bonaccorsi P, Calvieri S. Treatment of cutaneous and pulmonary sarcoidosis with thalidomide. *J Am Acad Dermatol* 1995;**32**:866–9
27 Brechtel B, Haas N, Henz BM, Kolde G. Allopurinol: a therapeutic alternative for disseminated cutaneous sarcoidosis. *Br J Dermatol* 1996;**135**:307–9
28 Henderson CA, Ilchyshyn A, Curry AR. Laryngeal and cutaneous sarcoidosis treated with methotrexate. *J R Soc Med* 1994;**87**:632–3
29 Lofgren S, Snellman B, Nordenstam H. Foreign body granulomas and sarcoidosis. *Acta Chirurgica Scandinavia* 1954; **108**:405–18
30 Hancock BW. Cutaneous sarcoidosis in blood donation venepuncture sites. *British Medical Journal* 1972;**4**:706–8
31 Iverson JMI, Cotterill JA, Wright V. Sarcoidosis presenting with multiple tattoo granulomata. *Postgraduate Medical Journal* 1975;**51**:670–2
32 Collins P, Evans AT, Gray W, Levison DA. Pulmonary sarcoidosis presenting as a granulomatous tattoo reaction. *British Journal of Dermatol* 1994;**130**:658–62
33 Savin JA. Sarcoidosis. In: Champion RH, Burton Jl, Ebling FJG, eds. *Rook Wilkinson Ebling Textbook of Dermatology,* 5th edn. Oxford: Blackwell Scientific Publications, 1992;**3**:2383–406

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