

CASE REPORT

Sarcoidosis Presenting as Acute Bilateral Parotid Swelling

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Sarcoidosis is a granulomatous disease of unknown origin usually affecting young and middle-aged adults.¹ The disease frequently presents with bilateral hilar lymphadenopathy, pulmonary infiltration, and ocular and skin lesions. Liver, spleen, lymph nodes other than hilar, heart, nervous system, muscles, bones, and other organs are less commonly involved.

The lungs are affected in more than 90 % of the patients with sarcoidosis and may present as hilar lymphadenopathy or parenchymal infiltration.¹ In later stages fibrosis with honeycombing, hilar retraction, bullae, cysts and emphysema may occur. Airways may sometimes be involved leading to airway obstruction and bronchiectasis. Parotid gland involvement is uncommon, occurring in less than 6% of patients.^{2,3} The parotid involvement is usually bilateral and the patient may present with swollen glands and dryness of mouth.

SUMMARY A case of bilateral parotid swelling of short duration, which turned out to be sarcoidosis confirmed by fine needle aspiration cytology, is presented here. The patient also had asymptomatic bilateral hilar lymphadenopathy and dryness of the eyes suggesting sarcoid involvement of the lungs and lacrimal glands. Sarcoidosis of the parotids, although rare, should be considered in the differential diagnosis of acute bilateral parotid swelling. Clinico-radiological findings of multi-organ involvement suggest the diagnosis in such cases.

Ocular involvement is relatively more frequent, occurring in 11 to 83% of patients with sarcoidosis⁴ and may present with dryness of eyes due to lacrimal gland involvement.

Sarcoidosis is rarely reported in India although the incidence of sarcoidosis is on the increase.⁵ The high prevalence of tuberculosis frequently obscures the recognition of sarcoidosis. A lack of extensive investigative facilities also hampers disease recognition. The pattern of organ involvement in sarcoidosis reported from India is similar to that in the west but sarcoid involvement of the parotid gland has been reported in one series

only.⁶ This prompted us to report a case of sarcoidosis of the parotid glands who initially presented with bilateral parotid swelling only.

CASE REPORT

A 49-year-old housewife was referred to our outpatient department for assessment. She had a ten day history of swelling at both angles of the jaw along with decreased salivation and loss of taste. She also gave a history of low-grade fever, malaise and dryness of the eyes for the same duration. There

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was no complaint related to the respiratory system. The patient had taken complete anti-tubercular treatment for cervical tubercular lymphadenopathy ten years earlier. She had undergone cholecystectomy eight years ago for cholelithiasis.

Her physical examination revealed bilateral firm non-tender swellings at both angles of the jaw. The remainder of her physical examination was unremarkable. A hemogram revealed a hemoglobin level of 12.9 g/dl, a total leucocyte count of 5,380 cells/mm³ with a normal differential count. ESR was 30 mm in the first hour (Westergren method). Urine examination was normal and fasting blood sugar was 73 mg/dl.

Fine needle aspiration cytology (FNAC) of the swellings had been performed on both sides and showed lymphoid cells, macrophages, epitheloid granulomas and Langerhan's giant cells and the pathologist suggested a diagnosis of tubercular lymphadenitis.

A chest radiograph (Fig. 1) was taken at presentation, which displayed bilateral hilar lymphadenopathy. The presence of bilateral hilar lymphadenopathy put the suggested pathological diagnosis in doubt. Moreover, the specimen slide had not shown any caseation or acid-fast bacilli on special stain. Biopsy of the parotid gland and bronchoscopy was suggested but the patient declined to undergo these procedures. Therefore, the FNAC slides (Fig. 2) were sent to another pathologist for review who reported it as discrete, compact, non-caseating epitheloid granulomas in salivary tissue consistent with sarcoidosis of

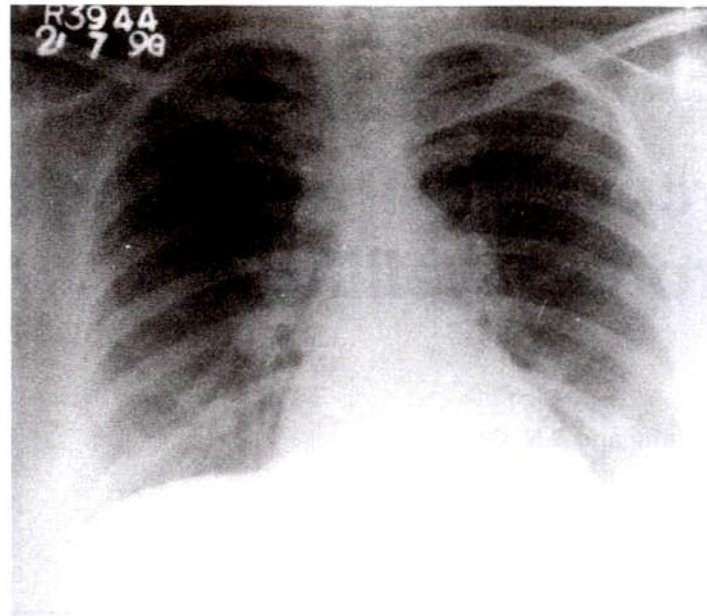


Fig. 1 Chest radiograph showing bilateral hilar lymphadenopathy.

the parotid gland. To confirm the diagnosis, the slides were reviewed by a third pathologist who also found non-caseating granular matter parotitis with giant cell reaction and suggested sarcoidosis as the diagnosis.

A Mantoux test did not raise any induration after 48 hours. Her serum calcium was 7.9 mg/dl (ref. 8.6-10.2); the 24-hour urinary calcium was 467.5 mg/d (ref. 100-200). Her serum angiotensin-converting enzyme (ACE) was 43 (ref. 8-52), rheumatoid factor was 124 (ref. 0-14) and IgG to A 60 *Mycobacterium* by ELISA was 76.3 (ref. 0-125). The LE cell phenomenon was negative and anti-DNA (double stranded) antibody was 3.6 (ref. 0-5.3). A complete lung function test and diffusion capacity were normal. The

ECG showed occasional ventricular ectopic beats, but within normal limits.

Contrast enhanced computed tomography of the thorax showed multiple lymph nodes in pretracheal, paratracheal, paraaortic, carinal and both hilar regions. Diffuse ground glass haze was noted. Subpleural and parenchymal reticulations and areas of fine nodularity were seen in both lung fields. Subpleural nodules and pleural tags were observed in the posterior segment of the right upper lobe.

The diagnosis of sarcoidosis of the lung, parotid and lacrimal glands was made. The patient was initially prescribed symptomatic treatment in the form of artificial tears. After four months follow up

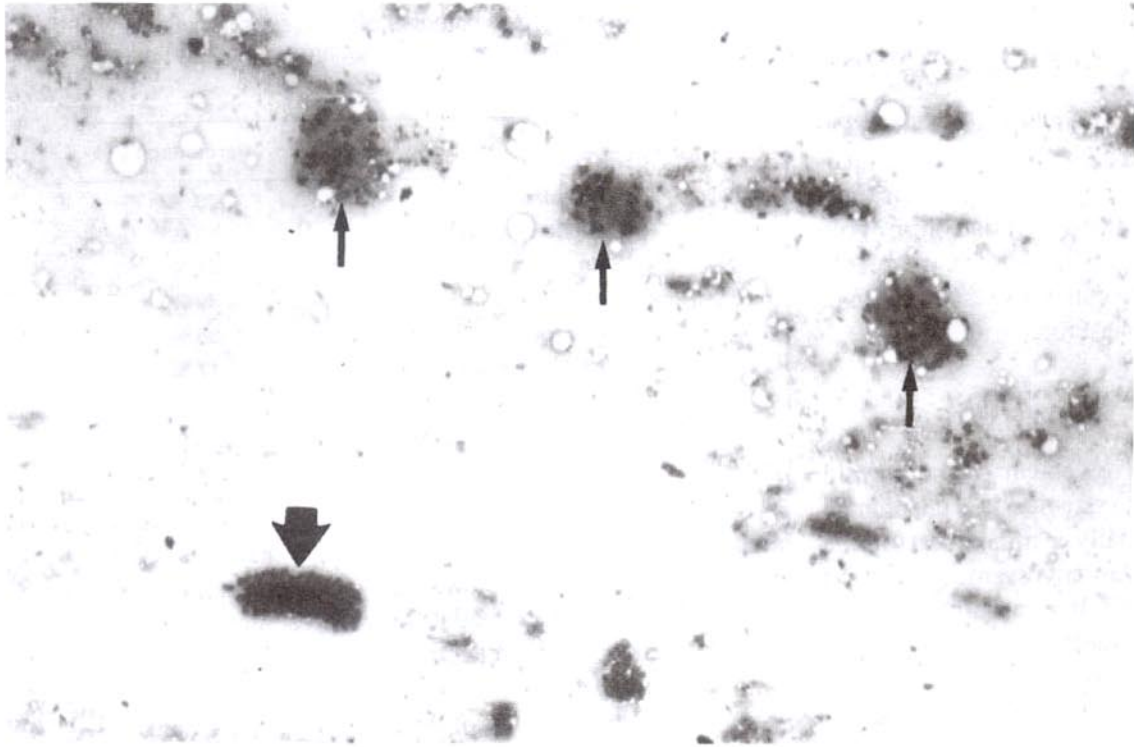


Fig. 2 Fine needle aspiration cytology showing multiple, discrete, compact, non-caseating epithelioid granulomas (thin arrows) along with a salivary duct (thick bold arrow).

the parotid swelling had subsided but the patient had developed intractable cough. She had also lost around 4 kg over this period. The serum ACE had risen to 300 U/l, but there was no significant change on the chest radiograph. The pulmonary function test had deteriorated with a fall of 350 ml in Forced Vital Capacity (FVC), Forced Expiratory Volume in the first second (FEV_1) had decreased by 600 ml and the new FEV_1/FVC ratio was 61%. At this stage the patient was initiated on prednisolone (40 mg/day), which was tapered gradually to 5 mg on alternate days. After 4 months of prednisolone treatment the patient became asymptomatic and radiological clearance was observed. The serum ACE had

Table 1 Involvement of various tissues in patients with sarcoidosis¹

System	Percentage
Lungs	>90 %
Peripheral lymph nodes	~33 %
Heart	~5 %
Liver	50-80 %
Skin	~25 %
Ocular lesions	11-83 %
Nervous	<10 %
Musculo-skeletal	25-39 %
Gastro-intestinal tract	<1 %
Hematological	4-40 %
Parotid glands	~6 %
Endocrine (hypercalcemia)	2-10 %
Genito-urinary	rare

fallen to 48 U/l and the 24-hour urinary calcium had dropped to 226 mg. The patient is still continuing the prednisolone treatment for over two years as she develops intractable cough every time the corticosteroids are discontinued. The dryness of the eyes has persisted but the patient is comfortable with the artificial tears. A recent pulmonary function test is normal, as is the chest radiograph.

DISCUSSION

Involvement of the parotid gland due to sarcoidosis is uncommon and is usually a minor part of the multi-system involvement seen in sarcoidosis.³ It is usually bilateral, slightly commoner in women, and presents mainly in the 20 to 40 year age group. There is widespread involvement of other systems, particularly intrathoracic. Patterns of involvement may sometimes be pathognomonic as in Heerfordt's syndrome (characterized by a chronic, febrile course, enlarged parotid glands, uveitis and facial nerve palsy).¹ Our patient, a 49-year-old woman, however, presented acutely with low-grade fever and bilateral painless swelling at the angles of the jaw. Moreover, she gave a history of treatment for cervical tubercular lymphadenopathy in the past, which made us suspect tuberculosis as the cause of these swellings. But a chest radiograph revealed bilateral hilar lymphadenopathy. This along with the absence of caseation and acid-fast bacilli in the fine needle aspiration cytology led us to question the initial FNAC diagnosis of tubercular lymphadenopathy. The slides were thereafter sent for review to two different pathologists and both reported it as

Table 2 Involvement of other tissues in patients with sarcoidosis and enlargement of the parotid glands²

System	Percentage
Intrathoracic	78 %
Peripheral lymph nodes	65 %
Skin	43 %
Spleen	39 %
Uveal tract	35 %
Lacrimal glands	17 %
Facial nerve	4 %

parotid sarcoidosis. A CT done later confirmed the hilar lymphadenopathy and demonstrated involvement of other mediastinal lymph nodes and lung parenchyma.

The simplest way of differentiating sarcoid parotitis from other causes of enlargement of the parotid gland is by seeking corollary evidence in other tissue systems, as was demonstrated in our patient. Although the most certain way of diagnosing sarcoidosis is by obtaining histological proof of sarcoid tissue through biopsy², fine needle aspiration cytology may be a reliable and cost effective method of diagnosis when used in conjunction with radiological and laboratory data,⁷ especially in patients unwilling to undergo the more invasive biopsy procedures. The presence of discrete, compact, non-caseating epithelioid granulomas in salivary tissue is an invariable feature of sarcoidosis of the parotid, as was seen in our patient.

In countries with high prevalence of tuberculosis, sarcoidosis may be misdiagnosed as tuberculosis as the two conditions occasional-

ly resemble each other in clinical and histo-cytological manifestations, as was the case initially in the report presented here. Thus clinician's diligence in pursuing evidence for sarcoidosis involvement of various organs is crucial for establishing an accurate diagnosis and the proper management.

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