An unusual case of sarcoidosis

By L. Vlahos, V. Benakis, K. Kotoulas and Gr. Pontifex

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From the Institute of Roentgenology, University of Athens, Areareion Hospital (Director: Professor Gr. Pontifex)

In the early stages of sarcoidosis the pulmonary as well as the constitutional symptoms are usually minimal or absent in spite of the marked radiological changes in the chest radiograph (Davidson and Macleod 1972, Krupp and Chatton 1972, Price 1973). The manifestation of sarcoidosis in the various organs rarely precedes the pulmonary hilar lymphadenopathy (Sommer 1963). On the other hand, in the early stage of sarcoidosis might show a tendency for generalized involvement (Uehlinger 1955). The typical pulmonary appearance of this disease (stage 1), as well known, is a bilateral hilar lymph node enlargement with or without paratracheal nodes involvement (Wurm 1960, Sutton 1975, Sommer 1975). This paper describes a case of sarcoidosis with unusual clinical and chest radiological presentation.

Case report

A 56 year old female, was admitted to our Hospital for the investigation of a fever of four weeks duration which was accompanied by rigor, night sweats, dry cough and loss of 5 kg of weight. The fever was reaching 37.8° C in the evening the first two weeks during which she was put by a physician an ampicillin with no apparent result. Chest radiograph 13 days after the onset of the illness did not show any significant abnormality. The temperature was rising steadily and by the end of the fourth week it was varying in the evening between 39.2° and 39.5° C.

The patient could not recall any serious ailment in the past, apart from an admission to another University Hospital in Athens in 1971 for the investigation of a hypochromic anaemia for which no apparent cause was found and was attributed to microscopic haemorrhage from a possible peptic ulcer.

On physical examination there was evidence of some mobile nodes in both sides of the neck as well as in the inguinal regions to which no particular attention was paid and an exanthema in the back having a linear distribution which was attributed to a variety of herpes.

Laboratory studies revealed a sedimentation rate of 85 mm in the first hour, a haemoglobin of 12 gr and a leucocyte count of 6,500 with normal distribution. The Mantoux reaction was considered as negative. Vidal and Wright reactions were also negative. The liver function tests were within normal limits. Urine analysis did not show any significant abnormality.

Repeat chest radiograph and tomograms the day of the admission i.e. two weeks after the first radiological examination revealed an ovoid mass in the right paratracheal region (Fig. 1). The right main bronchus and the demonstrated anterior and apical segmental bronchi were patent (the apical slightly narrowed). A malignant lymphoma, an
Fig. 1a A soft tissue mass is noted in the right paratracheal region.

Fig. 1b Tomography demonstrated an ovoid mass paratracheally. Calcification is also noted in the medial part of the mass.

Fig. 2 Moderate generalized enlargement of the inguinal and iliac lymph nodes on both sides with evidence of large filling defects. The shadow represents residual from a recent barium meal (arrow).
The described case of sarcoidosis was atypical firstly, because of its...

Discussion

Sarcoidosis in the early stages presents minimal or no symptoms at all despite the chest radiological changes, which could be quite alarming. The expression of the "healthy patient with the diseased chest" is appropriate for sarcoidosis. The described case of sarcoidosis was atypical firstly, because of its severe clinical picture and secondly of the unusual chest radiological appearance.

Clinical symptoms as high temperature and severe chest pain escape the usual pattern. It is also worth mentioning that the patient visited a general practitioner 9 months ago for some spots in her scalp which were thought to be due to the hair dye but according to the dermatologist it was possibly a manifestation of sarcoidosis.

Fig. 3 Histological image of an inguinal node section showing granulomatous changes typical of sarcoid.

Although there was no evidence of any previous illness the possibility of a recently started sarcoidosis is less likely because of the patient's age. The most likely explanation is that this was a recurrence of a disease which started either long before ad passed unnoticed or during the previous admission for the investigation of the sideropenic anaemia for which no apparent cause was found and which could explain the atypical chest radiological appearance which is not necessary to follow the usual pattern. It is also worth mentioning that the patient visited a general practitioner 9 months ago for some spots in her scalp which were thought to be due to the hair dye but according to the dermatologist it was possibly a manifestation of sarcoidosis.

References


Dr. L. Vlahos, D.M.R.D., D.M.R.T., F.R.C.R.
Ass. Prof. Dr. V. Benakis
Ass. Prof. Dr. K. Koutoulas
Professor Dr. Gr. Pontifex, Chairman
Institute of Roentgenology,
University of Athens, Arctaeon Hospital
76, Vass. Sophias Avenue, Athens, Greece

anaplastic bronchogenic carcinoma and a secondary deposit were considered as the most likely possibilities. Bronchoscopy failed to show any abnormality apart from slight rigidity of the lower end of the trachea. Sputum and scrapings from this part of the trachea did not show any malignant cells. Scalene node biopsy showed changes similar to those of sarcoid. This diagnosis was considered with great reservations because of the serious clinical picture to which by this time a severe central chest pain was added. A new chest radiograph and tomograms 8 days after the previous examination showed further enlargement of the paratracheal ovoid mass. Lymphangiography revealed generalised node enlargement in the pelvis and both groins with findings indicating possible lymphosarcoma or Hodgkins disease (Fig. 2). The paraortic chains were not filled. Biopsy of a right inguinal lymph node revealed again changes of sarcoid (Fig. 3).

Another look at the macular exanthema at the patients back by a specialist and excision of a macule confirmed again the diagnosis of sarcoidosis. The serum calcium was 12 mgr/100 nil. Nickerson-Kveim test was not performed.

Radiographies of both hands and feet were normal. The patient was put on 30 mgr prednisone daily. Four days later she had a spectacular regression of all the symptoms. The temperature went back to normal. Ten months later the patient is disease which started either long before ad passed unnoticed or during the previous admission for the investigation of the sideropenic anaemia for which no apparent cause was found and which could explain the atypical chest radiological appearance which is not necessary to follow the usual pattern. It is also worth mentioning that the patient visited a general practitioner 9 months ago for some spots in her scalp which were thought to be due to the hair dye but according to the dermatologist it was possibly a manifestation of sarcoidosis.

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