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Nodular lesions in the spleen diagnosed by US and CT in the course of sarcoidosis

Sarcoidosis is a systemic, granulomatous disorder with predominant involvement of hilar lymph nodes and pulmonary parenchyma. Extrapulmonary localization of sarcoidosis mostly concerns the skin and eyes, heart, central nervous system, kidneys, and very rarely the disease affects the liver and the spleen. Acute sarcoidosis is characterized by erythema nodosum, arthritis and fever and causes lymphadenopathy of hilar lymph nodes in the lung and has a good prognosis. Chronic disease involves interstitial changes in the pulmonary parenchyma and has no characteristic symptoms. Its onset is often asymptomatic, but has a disadvantageous course, which ends with respiratory failure (7,17).

The sarcoidosis diagnosis is based on histopatologic analysis of changes in the lung, lymph nodes or other affected organs. Additional but equally important value is shown by both observation of pathologic alterations within chest and abdomen (chest X-ray, US, CT, HR-CT) and analysis of the material from bronchoalveolar fluid).

CASE REPORT

A 54-year-old female patient is described who, in January 2002, reported to the hospital with pneumonia symptoms not disappearing after therapy with antibiotics. Similar episodes of pneumonia occurred in the last few years. Moreover, the patient suffered from polyarthritis with erythema nodosum, diagnosed in February 2000.

The patient was hospitalized in the Pulmonary Department from February 2002 because of weakness, decreased effort tolerance and chest-X-ray abnormalities. We did not notice any disturbances in blood morphology and biochemical examination of serum but the fallout amounted to 53/100 mm. In the sputum and bronchoaspirat we did not ascertain *Mycobacterium tuberculosis* and the tuberculin test with RT23 was 0 mm. Spirometry and blood gas examination was normal.

The bronchoscopy showed the mucosal vasal network and characteristic submucosal shields. The results of immunological analysis of bronchoalveolar lavage fluid were not typical of sarcoidosis: lymphocytes – 26%, macrophages – 58.9% and the CD4:CD8 ratio amounted to 3.05 with percentage of T-cells – 76.3% of all lymphocytes. Mucosal biopsy revealed epithelial and giant cells with astral bodies, which are typical of this disease. Sarcoidosis was diagnosed and therapy with prednisolone 40 mg/24 h was introduced. The symptoms disappeared very quickly.



Fig. 1. Splenomegaly visible on US. In the parenchyma – numerous poorly separated hypoechogenic foci, up to 15mm in diameter

US and CT findings. The radiological diagnosis was started with the thoracic X-ray, in which widened hilar shadows and moderately intensified parahilar micronodular lesions were found.

The US examinations were performed using Logic 500 Pro Series with a wide-band, 5MHz probe; the peripheral lymph nodes were evaluated with a 11MHz probe. The US findings revealed no enlarged or structurally changed peripheral lymph nodes. The abdominal US showed the heterogenic liver but not enlarged and without visible focal lesions. The spleen was enlarged to 14.5 cm in its max. dimension. In the parenchyma, numerous, poorly distinguished, hypoechogenic, round and oval foci 5-15 mm uniformly distributed in the whole organ were observed (Fig.1). The US follow-ups performed every four weeks during prednisolone treatment revealed a gradual decrease in the splenic size up to 12 cm. In the first US follow-up, the number and size of focal lesions were smaller and difficult to differentiate from the remaining parenchyma. The thoracic HR-CT and abdominal CT was carried out with Somaton DRH (Siemens) in dynamic scanning after the administration of the contrast bolus (80ml of Ultravist 300) and Light Speed Ultra (GE Medical Systems). The thoracic CT demonstrated the micronodular lesions localized along parahilar vessels and bronchi and slightly enlarged hilar nodes of both lungs. The expiratory scanning revealed small, air-trapping, lobulous lesions in the lower lobes. There were no features of fibrosis in the pulmonary parenchyma. The radiological picture of the chest was found to be typical of stage 2 sarcoidosis. Steroid therapy resulted in almost complete remission. The abdominal contrast-enhanced CT showed visible, numerous, hypodense nodules (about 30 H.U.), up to 15 mm in diameter localized in the enlarged spleen. However, these foci were difficult to identify at pre-contrast scanning. No hepatic lesions and enlarged lymph nodes were detected. In the first follow-up, the number and size of splenic nodules decreased and they were well separated (Fig. 2 and 3). The follow-up performed 2 months later showed faintly visible focal lesions, but after next 3 months the spleen was normal.



Fig. 2. Postcontrast scanning reveals numerous low attenuation nodular lesions and splenomegaly



Fig. 3. Reduced number and size of hypodense foci in the spleen following treatment

DISCUSSION

The first description of nodular sarcoidosis of the spleen detected at CT was reported by Mathieu et al. in 1986 (9). In the following years only single cases (1,10) or short series (1,4,14) were described, but the biggest worldwide compilation was published by Warshauer et al. (14–16) in 1994-1995. It presented the analysis of 32 cases of the focal splenic lesions in sarcoidosis collected on the basis of radiological documentation of seven university centers in the USA between 1984 and 1994. The radiographic images of all the cases were similar – numerous, small hypodense nodules, 5-15 mm in diameter, located quite uniformly in the parenchyma. The authors presenting analogous cases agree on morphology of the lesions at CT (3,6,9,11,13). The only exception is the report by Folz et al. (4), who additionally described

multiple small calcifications (seven cases) and a solitary bigger tumour. In the majority of literature cases, the symptoms of splenomegaly were observed – ranging from small to severe, in which the mass exceeded 2.5 kg (2,3,5,13-16). However, Warshauer et al. did not find any statistical correlation between the presence of focal lesions and severity of splenomegaly. In our patient, the spleen was slightly enlarged but detectable on palpation and its size was rapidly decreasing during treatment.

On the basis of the literature data, the frequency of concomitant splenomegaly and focal lesions can be assessed as about 70% (1,3,11,13-16). In more than a half of the cases reported by American authors, the focal lesions in the spleen were accompanied by similar nodules in the liver; enlarged lymph nodes of the abdominal cavity were observed in 76% of cases (15). The other authors report similar findings (1,11). In our case, the thoracic changes were the typical small nodules located mainly in the parahilar areas, radiologically evaluated as stage 2. The case presented by us belongs to those rare cases in which nodular lesions in the enlarged spleen were the only manifestations of abdominal sarcoidosis. The full radiologic evaluation of the treatment effects can be found only in a few reports (6,8,10). Biopsy of the spleen was not performed as the abdominal complaints were mild, laboratory results normal; additionally, the patient did not give her consent. Similar doubts concerning aggressive diagnostic procedures in such lesions were reported by Warshauer (15).

A few histopathologically confirmed cases of nodular sarcoidosis of the spleen described in literature usually showed the symptoms of hypersplenism and marked clinical symptoms or their diagnosis based on thoracic lesions was inconclusive (13,14). Britt et al. recommend the differential diagnosis of focal splenic lesions in the course of sarcoidosis, lymphoma, tuberculosis and mycosis (1). In our patient, neither palpation nor radiology revealed any enlarged lymph nodes typical of lymphoma; laboratory tests for tbe and mycosis were negative. Moreover, there was no focus, which was likely to result in metastatic lesions. Our presumptive diagnosis was confirmed by the gradually reducing spleen mass as well as decreasing number and size of nodules during the prednisone therapy. The value of CT with the contrast medium bolus in the radiological evaluation should be underlined. In our examinations the nodules were not visible on pre-contrast scanning, they became very distinct after the contrast administration as they were markedly attenuated compared to the remaining parenchyma. Their shape, size and location corresponded to the features described in literature (6,11,13–15).

In our center, the US examinations are routinely performed in the patients with sarcoidosis immediately after the thoracic lesions have been detected and regularly during observation and treatment. In the cases in which the abdominal changes were detected and the treatment was instituted, the follow-ups are performed every four-six weeks. US of our patient revealed the focal lesions and allowed us to evaluate the organ size; however, on the subseguent examination the nodules became less visible. The pulmonary lesions were decreasing with the regression of splenic changes and improvement of the clinical condition of the patient. It is interesting that the focal splenic lesions in sarcoidosis are so rarely detected although in biopsy of the spleen sarcoidal granulomata are found in 24–53% of the patients (11,12,15). Moreover, the incidence of lesions demonstrated on post mortem examinations is high, ranging from 40 to 80% (17).

In the reports published before US and CT became widely used, the data concerning the spleen lesions in the patients with sarcoidosis were scarse as the diagnosis was mainly based on the spleen enlargement and infrequent biopsies.

In a big clinical compilation involving 6074 cases of sarcoidosis, the frequency of splenomegaly was found to be about 10% (5). In the Polish studies published at that time, which analyzed 522 cases of sarcoidosis, this frequency was only 2.1% (8). In the next radiologic studies based on US and CT, the data were higher ranging from 25 to 60% (1,4,14); however, those studies included the results based only on the selected groups of patients (16–59 cases) with clinical symptoms.

Nakata suggests that the cause of bigger nodules detectable on diagnostic procedures is the formation of aggregates of small granulomas surrounded by the fibrous tissue. This

phenomenon may be analogous to the one observed in HRCT of the lungs, in which bigger nodules consist of aggregates of numerous, small foci (10,14).

Another cause is likely to be the fact that CT of the abdominal cavity is relatively rarely performed in the patients with sarcoidosis, particularly CT with contrast bolus. This examination is usually conducted in the cases of undetermined US diagnosis of the abdominal lymph nodes. The extremely rare focal lesions found in the abdominal cavity do not justify the routine CT of this area. However, the patients with lesions detected on US and those with abnormal blood results, even when the US abdominal image is normal should be subjected to CT. CT may also be used as a useful tool to evaluate the effects of steroid therapy in focal splenic lesions (8,14).

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SUMMARY

The paper presents rare micronudular lesions found at US and CT in a 54-year-old patient with the stage 2 pulmonary sarcoidosis. The lesions were in the form of numerous nodules, 5-15mm in diameter, quite uniformly distributed in the parenchyma of the enlarged spleen. On the Us examination, the focus echogeneity was reduced, likewise on CT, in which the density at postcontrast scanning was found to be about 30H.U. However, no enlarged lymph nodes of the abdominal cavity and hepatic lesions, which often accompany nodular sarcoidosis of the spleen were detected. During steroid therapy, the condition of the patient improved, the pulmonary lesions subsided and the number and size of splenic foci gradually decreased. The case was compared to the available literature data concerning this uncommon form of sarcoidosis.

Zmiany guzkowe w śledzionie stwierdzone w badaniu KT i USG w przebiegu sarkoidozy

W pracy przedstawiono obraz rzadkich zmian drobnoguzkowych, stwierdzonych w badaniu ultrasonograficznym i tomografii komputerowej u 54-letniej pacjentki z drugim stadium sarkoidozy płucnej. Zmiany miały postać licznych guzków o średnicy 5-15 mm, rozmieszczonych dość równomiernie w miąższu powiększonej śledziony. W badaniu USG echogeniczność ognisk była obniżona, podobnie w CT, gdzie gęstość w skaningu pokontrastowym wyniosła ok. 30 HU. Nie stwierdzono natomiast powiększenia węzłów chłonnych jamy brzusznej i zmian w wątrobie, towarzyszących często sarkoidozie guzkowej śledziony. W trakcie sterydoterapii obserwowano poprawę samopoczucia pacjentki oraz stopniowe cofanie się zmian płucnych i równoległe zmniejszenie liczby i wielkości ognisk w śledzionie. Omawiany przypadek przedstawiono na tle przeglądu dostępnego piśmiennictwa, dotyczącego tej rzadkiej postaci sarkoidozy.