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Case report

Imaging findings in Langerhans' cell histiocytosis of the liver and the spleen in an adult

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Abstract. We present a case of Langerhans' cell histiocytosis (LCH) of the liver and spleen in an adult. The imaging features are different from those in the few previously reported cases of individual organ involvement by LCH.

Key words: Langerhans' cell histiocytosis – Liver – Spleen – CT – Ultrasound

Introduction

The term Langerhans' cell histiocytosis (LCH), formerly known as histiocytosis X, refers to disease entities including solitary or multiple eosinophilic granuloma, Hand-Schüller-Christian disease, Letterer-Siwe disease, and cases that do not clearly fit into any of the above categories. This entities share common pathologic features, i. e., aggregation of mature eosinophils and typical histiocytes, also known as Langerhans' cells. In most cases LCH is a systemic involvement in childhood. One case of LCH forming a solitary nodule in the spleen has been documented as an incidental finding on necropsy [1]. Isolated liver involvement by LCH has been reported three times [2–4]. Langerhans' cell histiocytosis confined to the liver and spleen has not been previously reported. Sonographic, CT, and follow-up findings in an adult with isolated involvement of the liver and spleen are the subject of this report.

Case report

A 44-year-old male presented with weakness, pain in the right hypochondrium and left shoulder, a 5-kg weight loss in 1 month, and night sweats. Clinical examination was normal. Serum levels of alkaline phosphatase (226 U/L; nl < 220), lipase (246 U/L; nl < 190), and Y GT (106 U/L; nl < 49) were elevated.

Sonographic examination showed multiple, well-defined, rounded and oval lesions of decreased echogenicity within the liver, varying in diameter from 1 to 2.5 cm (Fig. 1 a).

Contrast-enhanced CT showed multiple, well-defined, rounded, and oval low-attenuation hepatic lesions with subtle ring enhancement with the same diameters as measured on ultrasound (Fig. 1 b). There was mild splenomegaly and three well-defined, low-attenuation lesions were noted in the spleen. The splenic lesions were similar in appearance to the hepatic lesions.

The CT scans of the thorax, brain, and pelvis were unremarkable. Initially, CT-guided liver biopsy showed an inflammatory infiltrate of histiocytes and eosinophils. On immunohistochemistry the histiocytes stained positively for S-100 protein. Eosinophilic granuloma was the provisional diagnosis. A wedge biopsy of the liver was performed laparoscopically. The biopsy specimen contained two nodules of inflammatory tissue with multiple eosinophils centrally and numerous histiocytes, with folded and grooved nuclei and inconspicuous nucleoli, in the periphery (Fig. 2). These findings, combined with the strong immunoreaction on S-100 protein of the histiocytes, confirmed the diagnosis of eosinophilic granuloma.

Treatment with corticosteroids was implemented and CT after 1 month showed a complete resolution of the splenic lesions and a dramatic reduction in the number and size of the hepatic lesions. A CT examination 1 month later was normal.

Discussion

In the three previously reported cases of LCH confined to the liver in adults [2–4], sonography showed hyperechoic, irregular, nodular lesions. In our patient, the liver lesions were hypoechoic and well defined.

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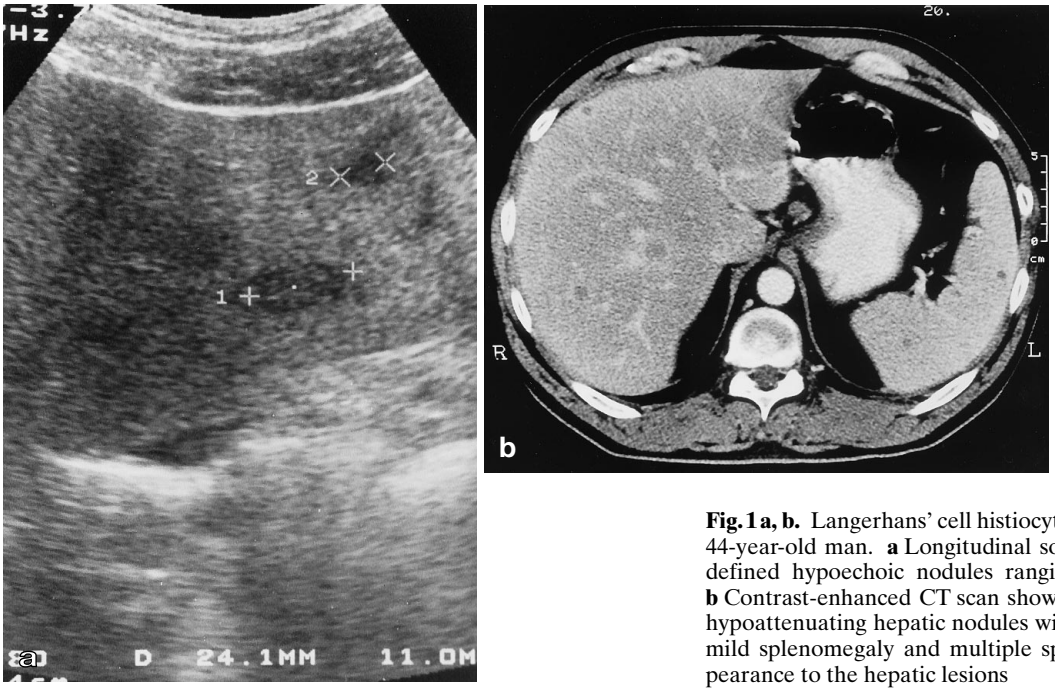

Fig. 1 a, b. Langerhans' cell histiocytosis of the liver and spleen in a 44-year-old man. **a** Longitudinal sonogram shows multiple, well-defined hypoechoic nodules ranging in size from 1 to 2.5 cm. **b** Contrast-enhanced CT scan shows multiple, well-circumscribed hypoattenuating hepatic nodules with ring enhancement. There is mild splenomegaly and multiple splenic lesions with similar appearance to the hepatic lesions

In the three previously reported cases CT showed hypoattenuating, irregular, nodular lesions. In our patient the lesions were also hypoattenuating and nodular but irregular lesions were not seen. The ring enhancement on contrast-enhanced CT, although unspecific, was also different from the previous reports where diffuse enhancement was described. A combination of features was also observed in one other case [2].

Regarding the similarity of the features on enhanced CT and equal response on corticosteroid therapy, we assume the splenic lesions also to be foci of LCH.

Combining the sonographic and CT findings, metastasis or granuloma could not be excluded.

Engelbreth-Holm et al. described the existence of four histological phases of LCH: proliferative, granulomatous, xanthomatous, and fibrous [5]. In that case xanthomatous lesions are likely to be hypoechoic and hypoattenuating; fibrous lesions, on the other hand, would be hyperechoic and hyperattenuating. The difference in echogenicity, delineation, and contrast enhancement of the lesions in comparison with previously reported imaging features might be due to a different histological stage of LCH lesions. Further investigation will be necessary to answer this question.

We conclude that, although very rare, LCH should be included in the differential diagnosis of widespread space-occupying lesions in liver and spleen.

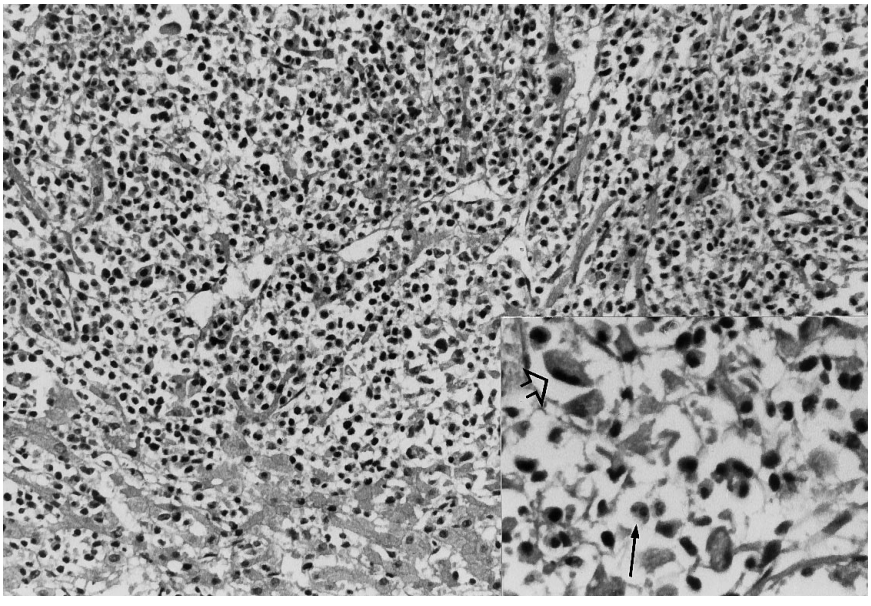

Fig. 2. Hematoxylin eosin safran liver section of the same patient showing Langerhans' cell histiocytosis (magnification × 25). The *inset* shows Langerhans' cells with typical nuclear grooves *(arrowhead)* and a number of eosinophils *(small arrow)* (magnification × 40)

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Book reviews

European Radiology

Martin-Duverneuil, N., Chiras, J.: Imagerie Maxillo-faciale [Maxillofacial imaging]. Paris: Flammarion Médecine-Sciences, 1997, 375 pp., 598 FF, ISBN 2-257-15553-X

This book is published in the series Imagerie Médicale (Medical Imaging), which aims to show the reader the possibilities of modern imaging, and to help the imaging specialist in choosing the most appropriate imaging method.

Many authors have contributed to *Imagerie Maxillo-faciale,* most of them French and some Belgian. As stated by the title, the book is devoted to imaging of the face, nose and paranasal sinuses, mandible and temporomandibular joint, and salivary glands. The work covers normal imaging findings, inflammatory, tumoral and traumatic pathology, and also gives some notions on congenital and vascular anomalies.

The very high quality of the images is apparent at first glance. There are several good textbooks on this subject, but this one also shows how imaging of the maxillofacial region should look: tailored to the problem, choosing the technical parameters so that maximal information can be derived from the images. In some practices, suboptimal or even poor imaging quality is delivered, including by people using sophisticated CT or MRI machines. This book shows how it should be done! Some of the figures were used previously in another recent French book on head and neck imaging.

Imagerie Maxillo-faciale offers the background information needed to choose the appropriate CT and/or MRI technique in a particular situation. The text is well written and to the point, and contains sufficient detail to come to a correct diagnosis or differential diagnosis in most cases. Well-structured tables and explanatory drawings of good quality have been included in several chapters.

The chapter on inflammatory nasosinusal pathology is interesting. Complications of sinusitis are nicely described and illustrated. The authors use the different types of inflammatory disease distribution, as described by Babbel and Harnsberger, to describe the CT findings in chronic sinusitis. In my opinion, this system is interesting from a educational point of view, but as typical examples of the different types are not often seen in daily practice, its application is not so easy.

I particularly liked the chapter on tumoral pathology. This contains a very interesting section on odontogenic tumoral and pseudotumoral pathology, a kind of pathology often treated rather superficially in other works on radiology of this region. In the non-odontogenic tumor section the authors describe quite extensively several rare lesions, while squamous cell carcinoma and adenocarcinoma are grouped, among others, under the title 'other malignant tumors of the maxillofacial region'. Personally I would have reserved more space for squamous cell carcinoma and adenocarcinoma, as these are amongst the more common malignant tumors of this region; a somewhat more detailed description (and illustration) of juxtatumoral meningeal enhancement and its meaning, as well of early brain invasion (mainly occurring at the anterior fossa), would have been interesting, as such findings may influence patient management. However, this is a personal opinion, and one can not give an exhaustive description of all possible occurrences in a limited volume.

In the chapter of maxillofacial trauma a thorough overview of imaging methods and findings in such patients is given. A minor criticism is that I do not think it is a good idea to try to visualize orbital fractures with three-dimensional (3D) reconstructions (figs. 6.1, 6.4), due to the artifacts associated with this technique. On the other hand, I think 3D imaging of the maxillofacial skeleton is very interesting in extensive and complex facial fractures (e. g. Le Fort type), where it helps by showing the displacement of fracture fragments and makes classification of the fractures easier. With spiral CT acquisitions, high-quality 3D images can be generated in most patients; this is mentioned in the text, but only one such example of 3D imaging in a complex trauma case is shown.

This book on maxillofacial imaging is of very high quality. As maxillofacial pathology is amongst the most common indications for head and neck imaging, I can recommend *Imagerie Maxillo-faciale* to all radiologists. Let us hope that an English edition will soon become available, so that dissemination of this work on a larger scale will be feasible. R. Hermans, Leuven

Markisz J. A., Ramirez de Arellano E. A., Kazam E., Rafal R. B.: MRI atlas of the chest: normal anatomy and pathology. Oxford: Blackwell Science, 1997, 306 pages, £ 95.00, ISBN 0-86542-391-1

The book contains 306 pages, printed on high-quality glossy paper. It is divided in four parts, all richly illustrated. Legends are clearly indicated. All these elements contribute to a pleasant appearance for the reader. Numerous references at the end of the book are useful to those who wish to study thoroughly the anatomy, principles of MRI and chest imaging.

The first three parts of the book detail anatomical structures of the chest in the standard axial, sagittal and coronal planes. The level of each anatomical section is well indicated, which contributes to the quality of the atlas. Such an atlas is very helpful in clinical practice. However, spatial resolution is poor.

The fourth part illustrates a wide range of the different entities that may involve the chest, including thoracic, cardiovascular, chest wall, breast and spine pathological cases. However, in some cases, the treatment of gadolinium enhancement may be open to criticism, partly due to the lack of analysis in the same planes before and after gadolinium administration. Moreover, some of the descriptions are insufficient or inadequate. Several of the cases could have been explored and diagnosed with spiral volumetric CT with longitudinal reformations.

In conclusion, this book may be of interest for radiologists who are not familiar with MRI of the chest and want to learn the technique. Despite the spatial resolution being of insufficient quality, such an anatomical atlas is very helpful in clinical practice. A wide range of various entities are shown, but it must be remembered that several of these cases could have been diagnosed with spiral volumetric CT. C. Beigelman, Paris