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* Compiled by Professor Willich, Heidelberg

above an area of narrowing should suggest a secondary pulsion diverticula above a foreign body. This is particularly true when the level of the neck of the diverticulum is above the thoracic inlet or the aortic arch, common foreign body sites.

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tern, marked edema and prominent mass effect and especially the "ring-like" peripheral pattern of enhancement. This peripheral rim was reported in only two previous cases of brain lymphoma of adult patients, one of them with AIDS [4, 9].

In conclusion, in childhood primary brain lymphoma is extremely rare [11, 12]. Our case presented atypical CT features, mimicking gliomas, or inflammatory disease, especially brain abscess. Despite its rarity brain lymphoma should be included in the differential diagnosis, even in the presence of unusual CT features.

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this high-risk group, Copel et al claim the four-chamber view has a positive predictive value of 95.8% and a negative predictive value of 99.4% [2]. Diagnosis of HLHS with 100% accuracy at 18–24 weeks gestation is accepted as the rule [1–5], although two atypical cases of HLHS have been described [4, 5] in which a left ventricle of initially normal size but poor contractility was noted during detailed fetal echocardiography.

Despite the high yield of fetal echocardiography in the high-risk group, most cases of major congenital heart disease will be found in the low-risk population group (2–3 per thousand). The four-chamber cardiac view, a standard part of the routine obstetric ultrasound examination [2, 3], is said to exclude most severe congenital heart disease (HLHS, atresia of the mitral, pulmonary or tricuspid valves, double inlet ventricle, atrioventricular canal defect) [2, 3], from 16 weeks gestation onwards. It has come to be expected that any competent obstetric ultrasound examination will detect HLHS and most (but not all) other severe congenital heart anomalies from 16 weeks gestation onwards, using the

four-chamber view alone. If cine-loop technology is not available, it is difficult at times to obtain an ideal four-chamber view on hard copy during a routine time-limited obstetric ultrasound examination, and some reliance is made inevitably on the real-time appearance seen by the sonographer.

The etiology of HLHS is unknown. The development of classical HLHS following less typical appearances in the second trimester in our case and the two atypical cases previously reported [4, 5] may indicate that the final expression of HLHS has a variable age of onset.

In the light of our case, we believe the following points can be made. HLHS cannot be confidently excluded by a normal four-chamber view in the second trimester. The accuracy of the four-chamber view in "routine" antenatal ultrasound is not well established, although the four-chamber view is reliable in diagnosing major congenital heart disease when it forms part of a detailed fetal echocardiographic examination. It is important for the sonographer to note function as well as structure when obtaining the four-chamber view.

Acknowledgement. We thank Dr. R. Chisholm for technical advice.

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(continued on p. 439)

curing in the anterior mediastinum [2]. To our knowledge our patient may be the first reported case presenting with prolonged fever due to infection and secondary abscess formation in the teratoma.

The most frequent tumors of this area in childhood are neuroblastoma and other neurogenic tumors, more rarely duplication of the gastrointestinal tract [2].

Diagnosis of postero-inferior mediastinal mass which was suspected on ultrasonography was confirmed by CT. Accurate localization and relations to

adjacent organs, together with confirmation of calcification and fat within the mass gave a likely diagnosis. Recognition of a probable abscess formation enabled the correct treatment to be established prior to operation.

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of mesenteric fibromatosis occurring in the absence of previous surgery [3].

A review of the Polyposis Registry at St Mark's Hospital, London revealed an association with thyroid carcinoma (particularly papillary carcinoma in women under 35 years) and recommended regular thyroid examination in all patients with adenomatous polyps [4]. Turcot's syndrome was originally described in 1959 with a report of two siblings with familial adenomatous polyposis one of whom had an associated medulloblastoma of the spinal cord, the other a glioblastoma of the frontal lobe. It has been suggested that the name Turcot's syndrome should be reserved for the association of FAP with medulloblastoma and

glioblastoma only, and that other cases of association with astrocytoma, neurofibroma, craniopharyngioma etc. are not true examples of Turcot's syndrome [5]. Our case is the first report of familial adenomatous polyposis in association with rhabdomyosarcoma.

Acknowledgements. We wish to acknowledge the help of Dr. B. Sandhu, Consultant Pediatrician.

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Blood cultures for bacteria, fungi and viruses were all negative. Serology for viruses was negative. Several bone biopsies revealed only non-specific inflammatory changes. Immunological studies were normal. X-rays revealed metaphyseal lucencies with adjacent periosteal elevations. Pathological fractures were noted at the distal ulna of the left arm (Figs. 1, 2). The axial skeleton, spine, pelvis and skull, were never affected. During a period of ten months, although she received several antibiotics, aspirin and steroids, swelling of the limbs remained unchanged.

X-rays of her limbs at the age of 1 year demonstrated tremendous bone changes. There was loss of normal bone modeling with extensive periosteal overgrowth (Figs. 3, 4). Repeat bone biopsy with electromicroscopy study showed distinct particles in the mitochondria, compatible with viral inclusions. Bone-tissue cultures for viruses were negative.

Anti-viral therapy with Acyclovir, and later Interferon did not have any beneficial result.

During four years of follow-up, no remission has occurred. The child remains crippled and bed-ridden, although her mental development is normal.

Discussion

The clinical, radiological and histological findings in the above case are compatible with a diagnosis of chronic recurrent multifocal osteomyelitis. However,

the excessive severe periosteal overgrowth is similar to the tumorous osteomyelitis published by Kozłowski [4]. Differential diagnosis includes those diseases with periosteal hypertrophy like Caffey's disease, Goldbloom's syndrome, diffuse sclerosing osteomyelitis (Garre's disease) and hyperostosis hyperphosphatemia syndrome.

The patient's disease started with metaphyseal destructive lesions, which are not a manifestation of Caffey's disease or Garre's osteomyelitis. She did not have any dysproteinemia or hyperphosphatemia to support the other entities [5, 6].

In the reported cases, no causative infective agent was ever found, yet Kozłowski et al. [3] and Speer [7] consider CRMO to be an infectious disease. We believe that the electromicroscopic findings suggest a viral etiology. The severe limb deformity, including overgrowth of the metacarpals and fingers, resulted in a crippled, bed ridden child, which is unusual in CRMO.

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