Segmental atrophy of the liver in a child: Case report and review of the literature

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Segmental atrophy of the liver (SAL) is a pseudotumour that is extremely rare in children. We report the case of a 4-year-old child who presented with abdominal pain and a non-tender mass at the epigastrium. A computed tomography (CT) scan revealed a complex lesion of the left lobe of the liver. At laparotomy, a mass arising from the caudate lobe was completely excised and the pathology report confirmed SAL. Although rare in children, paediatricians, surgeons and radiologists should consider the possibility of SAL in the differential diagnosis of hepatic masses in this group of patients.


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poorly circumscribed area of whitish fibrous tissue intermixed with semisolid gelatinous tissue. The surrounding liver parenchyma had areas of dark yellow tissue intermixed with similar white fibrous tissue. Microscopic examination revealed areas of elastosis to fibrosis and associated small islets of hepatocytes. At the periphery, there was liver parenchyma with fibrous tracts showing mild bile duct proliferation. Thin-walled veins were present in the fibrous septa. In the areas of fibrosis, there were a few vessels with prominent walls. The features were compatible with segmental atrophy of the liver (SAL), with no evidence of other pathological entities. The patient remains well 6 months after the operation. A follow-up ultrasound done three months after the surgery did not show any evidence of residual/recurrent disease.

Discussion

SAL is an under-recognised benign lesion. Though it is widely reported in adults, our review of the literature found only one paediatric patient: a 14-year-old. The lesions are usually much smaller than anatomical segments and often involve only a few hepatic lobules. Bile leakage from transected ducts resulting in inflammation and fibrosis to the small peribiliary capillaries may cause SAL. Larger SAL has been considered a complication of different benign and malignant diseases of the liver and of the bile ducts, including hydatid disease, cholangiocarcinoma, alcoholic cirrhosis, chronic active hepatitis with cirrhosis, hepatocellular carcinoma, cryptogenic cirrhosis, pyogenic cholangitis, sclerosing cholangitis, and acute hepatic failure. Benign stricture causes atrophy by biliary obstruction, while cholangiocarcinoma causes atrophy by biliary obstruction and/or portal vein branch compromise. Histology remains the gold standard of diagnosis.

Garg et al in their 6-case series reported that all 6 patients were female with a mean age of 58.3 years (range 37 - 80) and the mean size of the lesion 1.8 cm (range 3 - 3.6 cm). Singh et al in their 18-case series found that the median age of presentation was 63 years (range 14 - 91), female patients constituting 72% of the cases; 78% of the cases presented with right upper quadrant abdominal pain, the size of the tumour ranged from 1.8 to 10 cm and 83% of the lesions were subcapsular.

Our case of a 4-year-old girl is the youngest patient described in the literature. She presented with abdominal pain and a palpable mass. The lesion was located in the caudate lobe and it measured 9.5 cm × 7 cm × 2.5 cm. There is no specific laboratory test or biomarker for SAL. The histological features of SAL seem to vary based on when the biopsy is taken: early in the course (first stage), the lesion is characterised by collapsed hepatic parenchyma with associated islands of residual hepatocytes, chronic inflammation and marked bile duct proliferation. Later in the course (second stage), histological findings are characterised by little to no ductal proliferation, a decrease of chronic inflammation and increased amount of elastosis. As time passes and later in the course (third stage), histological findings show that the lesion is almost entirely composed of elastic fibres with scattered small islands of residual hepatocytes and portal tracts. In its latest stages (fourth stage), the lesion forms nodules with dense fibrosis. Based on the histology in our patient, she seems to have presented in the second stage. As demonstrated in our case, complete excision remains the treatment of choice.

Conclusion

SAL is a focal hepatic lesion that can mimic primary hepatic or metastatic lesions. It may present with abdominal pain and a mass. As demonstrated in our patient, paediatricians, surgeons and radiologists should be aware of this very rare condition, even in a child as young as 4 years of age. Histology remains the gold standard of diagnosis.

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References


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