Telangiectatic Focal Nodular Hyperplasia of the Liver in an Infant with Spontaneous Regression: A Case Report

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A distinctive mass in the liver in a two-month-old girl with elevated serum alpha-fetoprotein (AFP) level was diagnosed as telangiectatic focal nodular hyperplasia (FNH) after biopsy. The tumor spontaneously regressed and finally became no longer detectable by any imaging study within normal range of AFP. The nature of this novel entity and its management are discussed based on literature review.

Key words: telangiectatic focal nodular hyperplasia (FNH), infant, liver tumor, treatment, alpha-fetoprotein (AFP)

INTRODUCTION

Primary neoplasm of the liver in children can warrant assuming it as malignant hepatoblastoma or hepatocellular carcinoma [1]. However, there are other lesions including focal nodular hyperplasia (FNH), which can be diagnosed at any age but is usually diagnosed between 2 and 5 years of age in children [2–3]. Recently an entity of telangiectatic FNH was proposed and has been accepted as a new variant of benign hepatic lesion [7]. This paper reports an infant with a distinctive mass in the liver which had been diagnosed as telangiectatic FNH after biopsy but disappeared within two years. The nature of this novel entity and its management are discussed based on literature review.

CASE REPORT

A two-month-old female infant, who was born at 38 weeks of gestational age with 3070g of weight, with poor weight gain and frequent vomiting was referred to our hospital after she had been found to have a tumor-like lesion in her right hepatic lobe by ultrasonography. Imaging studies revealed that the lesion was 5 × 3 cm in size and was distinctively demonstrated as a mass. The mass was hypoechoinc in ultrasonography (Fig. 1a), and low density in the plain CT. The lesion was demonstrated as hypointensity in T1-weighted MR images and as hyperintensity in T2-weighted MR images (Fig. 2a). Her serum alpha-fetoprotein (AFP) level was elevated with 27,210 ng/ml (normal range at 2 months; 60–1100 ng/ml) [10]. Open biopsy was done with intent to start chemotherapy with preoperative diagnosis to be hepatoblastoma. However, the tumor was not pathologically confirmed to be malignant and was diagnosed as telangiectatic FNH (Fig. 3a, b). Careful follow-up imaging studies as well as serial AFP measurements revealed constant tumor shrinkage with AFP decline and finally no longer detectable by any imaging study within normal range of AFP by two years of age.

The tumor gradually became difficult to be demonstrated by ultrasonography within 2 years (Fig. 1b).

DISCUSSION

Telangiectatic FNH of the liver has been recently described as a new variant of FNH and Nguyen *et al.* reported that it accounts for about 15% cases of FNH [3]. Unlike classical FNH it lacks central scar and is more heterogenous, hyperintensity on T1-weighted MR images, strong hyperintensity on T2-weighted MR images, and persistent contrast enhancement on delayed contrast-enhanced CT or T1-weighted MR images [4]. It has been discussed where this new entity is situated within the spectrum of benign hepatic lesion. Recent genomic and protein analysis of the disease revealed that it is closer to hepatocellular adenoma rather than to FNH and the authors suggest it might have malignant potential [5].

Although classical FNH has been known to be found in pediatric age, telangiectatic FNH has been described only in 4 infants in the literature two each from Japan and Korea [7–9]. The initial case was described as a congenital hepatic tumor in a female newborn autopsied 4 hr after birth [7]. The three other cases were found in the resected specimens from infants whose preoperative diagnosis included hepatoblastoma [8–9]. Our case was managed without resection and followed up by regular imaging studies as well as AFP levels which resulted in spontaneous regression. Above mentioned cellular nature of the tumor may indicate its malignant potential but our experience would suggest this entity to be more benign nature [6]. Further case accumulation is necessary to elucidate the optimal management of this rare lesion but one should be conscious of the entity when a mass

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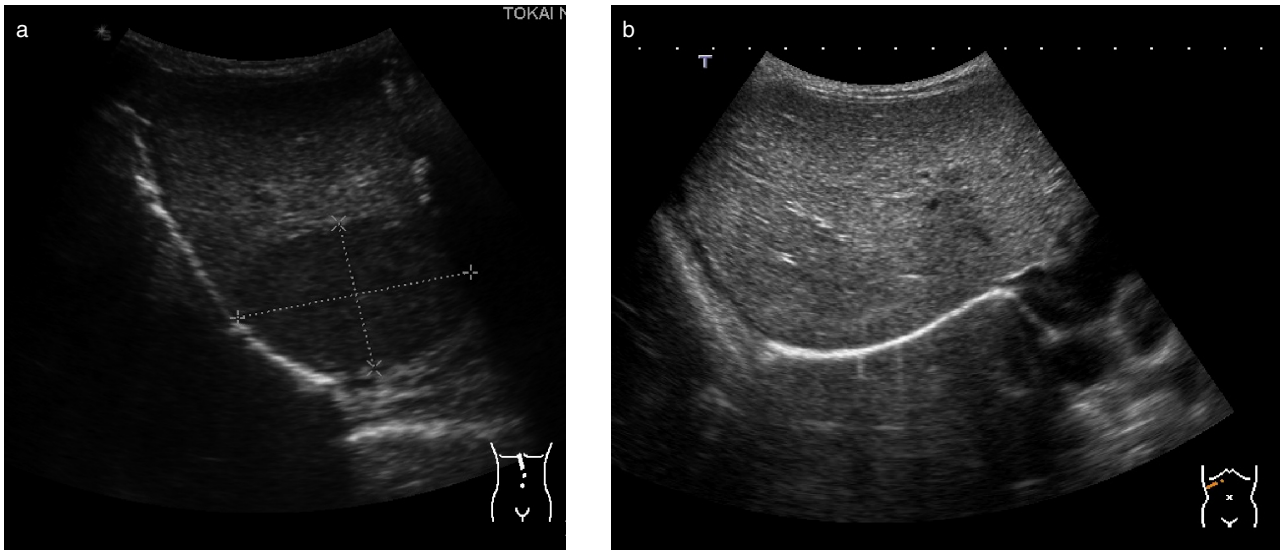

Fig. 1 Ultrasonographic appearance of the tumor at 2 months of age (a) and at 2 years of age (b)
a: The hypoechoic mass was seen in her right hepatic lobe. The leision was 5 × 3 cm in size and the margin was clear from normal liver.
b: The mass is not detected in her right hepatic lobe.

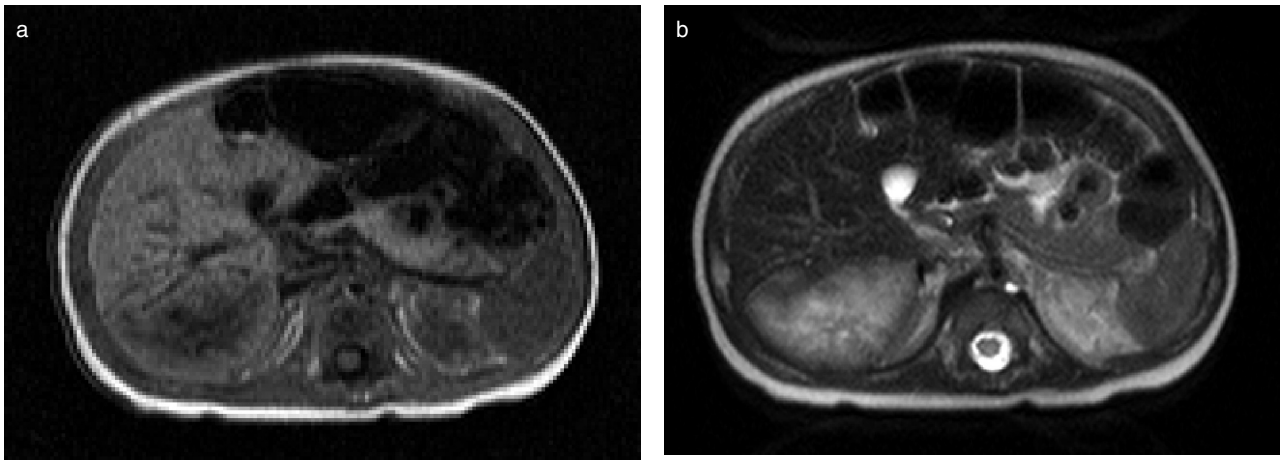

Fig. 2 MR images at 2 months of age
a: The leision was demonstrated as hypointensity at posterior lobe of liver in T1-weighted MR images.
b: The leision was demonstrated as hyperintensity in T2-weighted MR images.

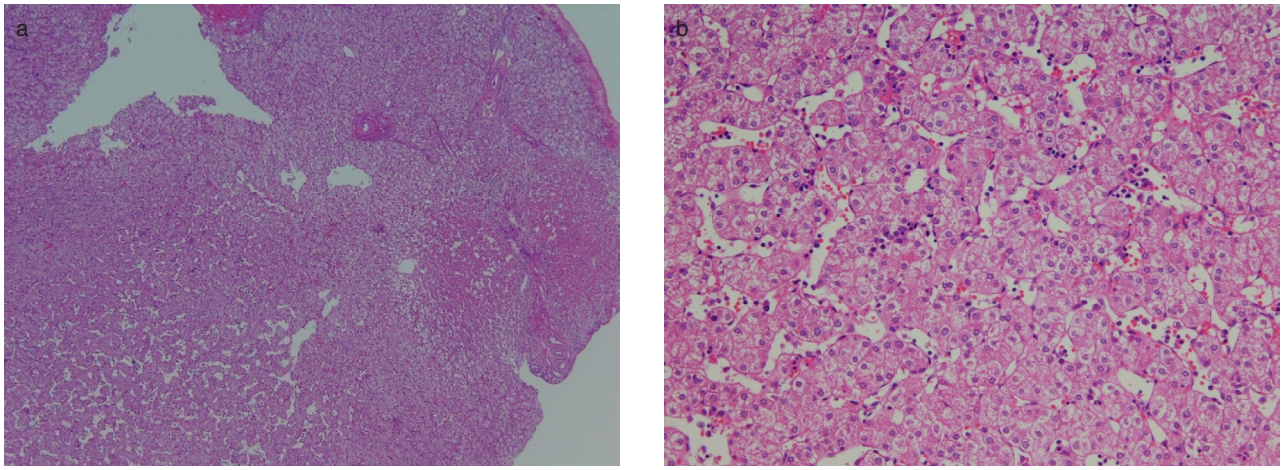

Fig. 3 Histopathology of the biopsied specimen
a: Pathologically, the tumor-like area (right-lower) was not clearly demonstrated from the normal hepatic tissue (left-upper).
b: In the tumor-like area sinusoidal dilatation is noted which is the characteristic feature of t-FNH.

Table 1 Reported infantile cases with telangiectatic FNH

Case	year	Author	Age	Sex	Preoperative Diagnosis	Location Size	Treatment
1	1974	Okayasu	stillborn	F	—	Right Egg size	Autopsy
2	2003	Kim	11 mo	M	Hepatoblastoma	Right 16 × 12 cm	Chemotherapy Resection
3	2005	Okamura	2 mo	M	Hepatoblastoma Endothelioma	Right 6.5 × 6 cm	Resection
4	2007	Kang	37 weeks	F	unknown	Right 35 × 3 cm	Resection
5	2008	present	2 mo	F	Hepatoblastoma Endothelioma	Right 5 × 3 cm	Biopsy

is detected in the liver in an infant.

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