## Focal Nodular Hyperplasia of the Liver

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Özet: KARACİĞERİN FOKAL NODÜLER Hİ-PERPLAZİSİ

Pek az sayıda cerrah ve patoloğun FNH hakkında kişisel deneyimi vardır ve hala hastalığın etiyoloji, patoloji ve tedavisi hakkında tartışmalar devam etmektedir. Buna karşın FNH, hakkında pek çok genelleme yapılmış olan nadir cerrahi antitelerden biridir.

Hastalığın sıradan özelliklerine uymayan 2 FNH'lı olgu sunulmaktadır. Görüleceği gibi her iki hastanın bulguları da FNH için sıradışıdır. Patolojik inceleme yapılana dek doğru tanı her iki hastayada konulamamıştır. Bu nedenle olguların orijinal ve ilginç olduğunu düşünüyoruz.

Anahtar Kelimeler: Fokal Nodüler Hiperplazi, Karaciğer

**F** ocal nodülar hyperplasia (FNH) of the liver and hepatocytic adenomas have received considerable attention since the initial report by Baum et al (1) in 1973, for the possible association between the development of these two lesions and oral contraceptives. Until recently, there wasn't a uniform system for pathological classification and trems applied for both of these entities were creating some confusion. Some of the historical synonyms for FNH are: hepatic hamartoma, parenchymal hamartoma, hamartomatous cholangiohepatoma, focal nodular cirrhosis, solitary hyperplastic nodule, liver cell adenoma and benign hepatoma (2). **Summary:** Few surgeons and pathologists have had much ersonal experience with FNH and still. Controversy exists about the ethiology, pathology and correct management. Un the contrary. FNH is one of the few surgical entites for which quite many generalizations have been made.

We present two cases with FNH which do not fit the ordinary criteria of the disease. As will be seen, findings in both of the patients were extraordinary for FNH. A correct diagnosis was not possible until pathological examination. Therefore, we believe that the cases are original, and interesting.

Key Words: Focal Nodular Hyperplasia, Liver

Few surgeons and pathologists have had much personal experience with FNH and still, controversy exists about the ethiology, pathology and correct management.

FNH is a benign liver tumor which is seen more frequently in women than men. The role of oral contraceptives in the ethiology is a possibility. It is encountered more often in the fourth decade. The disease is generally asymptomatic and is recognized accidentally during laparotomy or during evaluation of a non-related symptom. It prefers the right lobe and is between 1 cm. to 20cm. in magnitude. The presence of a central stellate scar in radiologic examinations is a characteristic finding (2,3,4,5).

One can hardly make such generalizations for only few entities in surgery. Besides, generali-

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zations may not be valid for every individual case. Recently we have operated on two cases which do not fit the ordinary criteria of FNH in our department. One of the patients was a 17 year old male, while the other was a 55 year old female who had never used oral contraceptives. Symptoms of the male patient were pain in the upper right quadrant for 3 years, with the addition of fatigue, weight loss, fever and mass in the upper right quadrant during the last month. Pain in the upper right quadrant was the only symptom in the female patient and had increased during the last 2 months. Both of the patients had had a Hepatitis A infection. Physical examinations revealed painful hepatomegaly in both of the patients and the male patient had an additional symptom of fever of 38°C. Blood, urine and stool examinations were normal. Also, the Casoni-Weinberg test,  $\alpha$ -fetoprotein and CEA were all in the normal range. The gastrointestinal system was normal endoscopically and radiologically. Ultrasonographical examination of the liver revealed: multiple solid masses in the medial and left hepatic lobe in the male patient and a solitary solid lesion about 50mm. in the left hepatic lobe in the female patient. a solid tumoral mass, completely occupying the left hepatic lobe in the male patient and a heterogenous benign lesion extending medially in the left hepatic lobe of the female patient were observed in CT. Scintigraphic examinations confirmed the CT findings. The male patient was also examined angiographically, and the findings were suggestive of a malignancy. Fine needle aspiration biopsy of the liver was made in the female patient and the histopathological examination revealed a benign lesion. It was decided to follow the female patient but to operate the male patient because of suspected malignancy. There were no pathological findings except for a tumoral mass in the lateral segment of the left hepatic lobe during laparotomy. Left hepatectomy was performed. The pathological diagnosis was FNH. The postoperative period was uncomplicated and the patient was discharged in the 7th postoperative day. 9 month

later, the patient had recovered completely and was free of his symptoms. The female patient was seen a month later. The symptoms persisted. Ultrasonografically, an enlargement of the lesion in her left hepatic lobe was observed. Left lateral segmentectomy was performed. During laparotomy, the only pathological finding was a lesion in the left hepatic lobe. The histopathological diagnosis was FNH. The patient was discharged from the hospital in the 9th postoperative day, after an uneventful postoperative period. She was relieved of all her symptoms on follow up, 7 months after surgery. Findings in both of the patients were extraordinary for FNH, a diagnosis was not possible until pathological examination.

## DISCUSSION

The ethiology of FNH is not clear. There have been many reports on its association with oral contraceptives. But the difficulty about building up this association lies in the fact that the incidence of FNH before worldwide usage of oral contraceptives is unknown. Also, from time to time, the disease is encountered in men and children (2). From another point of view, FNH is te hyperplastic response of the parenchyma to a developmental vascular malformation (6). There is also some evidence of its development following portoenterostomy in children (7). Hepatitis A infection in the history of both of our cases is an interesting finding.

Bleeding is a rare (2.5%), but most important complication (5). FNH dosen't have potential for malignancy but it may mimic a malignancy. It is usually impossible to differentiate FNH from a malignant lesion without a histopathological examination.

Goldin and Rose (8), have reported a case of multiple FNH of the liver with central nervous system neoplasia and vascular malformation which Wainless had previously defined. We also believe that it is mandatory to evaluate the CNS when FNH is suspected.

On gross examination, FNH is uncapsulated but sharply demarcated from the surrounding tissue. On cross-section, septa radiating from the central fibrous scar form nodules in the tumoral mass, which is pathognomonic for FNH. There is neither necrosis nor haemhorrhage (2,3).

Microscopically, normal hepatocytes form cirrhosis-like nodules around the centrally located bile ducts. There are also Kuppfer cells and vascularization of the central scar to some extent. On the other hand, in hepatocytic adenomas there is hepatocytic proliferation without bile-duct proliferation (9). Radiological examinations are usually helpful but a diagnosis cannot be established solely through x-ray studies.

USG is sensitive but not specific for FNH (5).

The hypodense image in CT on the precontrast scan becomes hyperdense with IV contrast. But rarely it is isodense during both of the imaging periods. In 9% of the cases, the CT image cannot be retained. The pathognomonic central stellate hypodence scar can be seen in only 23% of the cases. The presence of haemorrhage is suggestive of an adenoma (5,7).

Angiography displays a hypervascular image and a dense capillary stain. Atypically, the lesion is sometimes avascular. A characteristic feature, unique to FNH is the demonstration of central stellate septations which are encountered only in 29% of the cases (5).

"Hot spot" appearance in radionuclide imaging represents bile duct proliferation. But it must be kept in mind that the same appearance can also be seen in Budd-Chiari syndrome, vena cava superior syndrome and tricuspid infufficiency. Colloid uptake in colloid scintigraphy is usually normal or decreased. Very few patients display an increased colloid uptake (10,11,12). The characteristic central scar is also noted in Magnetic Resonans. Especially in T2 weighted sequences, a hyperdense scar is typical, but not specific. It is differentiated from primary malignant hepatic tumors by the homogeneity of the tissue around the central scar and the uniformity of the signal intensity in the normal parenchyma and the tumoral tissue (13,14).

Fine needle aspiration biopsy can be useful in the diagnosis of FNH but not in every case. Surgical biopsy should be prefered if an adenoma is suspected (9).

The incidence of other benign or malignant tumors in patients with FNH is not rare (13). Hepatic hemangioma is observed in 23% of FNH cases (15).

Generally, if FNH is diagnosed accidentally and the patients are asymtomatic, they should be followed-up non-operatively. CT is useful in follow-up. Patients with FNH should quit using estrogens. Surgical intervention is necessary if enlargement of the lesion is noted.

Ethanol embolotherapy is sometimes helpful in the management of FNH. The basis for this approach is that FNH is supplied by a solitary artery and there are no A-V shunts in the tumoral tissue (16). We have had no experience with this procedure.

As a rule of thumb; symtomatic FNH should be resected.

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