A rare malignancy in a patient with neurofibromatosis: gastric adenocarcinoma

Bora Barut,1 Volkan Ince,1 Fatih Ozdemir,1 Murat Harputluoglu,2 Burak Isik1
1 Department of General Surgery, Inonu University Faculty of Medicine, Malatya, Turkey
2 Department of Gastroenterology, Inonu University Faculty of Medicine, Malatya, Turkey

ABSTRACT

Neurofibromatosis is an autosomal dominant disease that is characterized by cutaneous hyperpigmentation and multiple neurofibromas and often accompanied by localized peripheral and central neural tumors. Gastrointestinal involvement is seen in 25% of the cases. Gastrointestinal stromal tumors, pancreatic and duodenal neuroendocrine tumors are the most common tumors in patients with neurofibromatosis. Neurofibromatosis and gastric adenocarcinoma constitute an extremely rare combination. In this case report we aim to present a patient with neurofibromatosis operated due to gastric adenocarcinoma.

Keywords: Adenocarcinoma; neurofibromatosis; stomach.

CASE REPORT

In a 67-year-old male patient, upper gastrointestinal endoscopy performed upon complaint of the patient revealed a polypoid mass extending from gastric corpus up to antrum which was histopathologically diagnosed upon examination of the biopsy specimen excised from this mass lesion. With café-au-lait spots detected all over his body (Figure 1A), and neurofibromas (Figure 1B) the patient was evaluated in collaboration with the department of dermatology, and the diagnosis of neurofibromatosis was made. On examinations performed by Departments of Eye Diseases, and Neurosurgery, any involvement of eye, and brain could not be detect-
ed. On abdominal computed-tomograms, a 6 cm-
polyoid mass localized on the greater curvature of
the stomach, and extending up to the lumen, and
a nearly 1 cm-long hypodense lesion in the 8th he-
patic segment were observed (Figure 2). This lesion
on the 8. hepatic segment was rather evaluated by
radiologists as a simple cyst. Therefore, any preop-
erative intervention targeted at this lesion was not
considered, and perioperative excisional biopsy was
decided on.

At admission, his whole blood counts, and bio-
chemical parameters were within normal limits.
Only CA 19–9 level was increased (48.3 U/ml
[normal 0–33 U/ml]).

During laparotomy, a mass lesion extending be-

\[ \text{Figure 1. (A) Café-au-lait spots. (B) Neurofibromas.} \]

\[ \text{Figure 2. Computed tomography of the patient.} \]

\[ \text{Barut et al., Gastric adenocarcinoma} \]
During postoperative period the patient is currently being followed up by the departments of eye diseases, and neurosurgery regarding the presence of neurofibromatosis. Department of dermatology initiated medical treatment. In addition, eye, and brain involvement were not encountered in examinations, and tests. The patient is being followed up without detection of any problem.

DISCUSSION

Neurofibromatosis Type I (von Recklinghausen disease) was firstly diagnosed in the year 1882. It is an autosomal dominant genetic disease seen in every 3000 births which develops secondary to mutation on the chromosome 17q11.12. In this disease newly formed mutations are frequently seen with an equal distribution between male, and female patients [1, 4].

In association with neurofibromatosis, neurogenic, and endocrine tumors as meningioma, glioma, pheochromacytoma can be seen in 3–5% of the cases [5]. In a few cases concomitancy between neurofibromatosis, and gastric adenocarcinoma has been reported [6]. In neurofibromatosis patients with gastrointestinal tumors signs, and symptoms as abdominal pain, bleeding, and manifestations of intussusception are present, and most of the cases are incidentally detected during laparotomies performed for other indications, postmortem examinations, and gastrointestinal barium series [7]. In patients with neurofibromatosis the most frequently seen gastrointestinal tumors are stromal tumors which frequently localized on stomach, and small bowels [4].

Conclusion

Since gastrointestinal tumors in patients with neurofibromatosis do not yield many specific symptoms, diagnosis of this disease is usually delayed. In fact, our patient who had not any complaint other than hematemesis, the diagnosis of gastric adenocarcinoma could be made only following endoscopic examinations. To prevent delays in diagnosis, patients with the established diagnosis of neurofibromatosis should be examined as for the presence of gastrointestinal malignancies, and apart from gastrointestinal stromal tumors, diagnosis of gastric adenocarcinoma should be kept in mind.

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