Signet ring cell carcinoma of the ampulla of Vater is extremely rare. The 7 cases reported earlier have been in older patients. We report a 32-year-old lady with this condition, who also had metastases in the bone marrow, vertebrae, lungs and liver. [Indian J Gastroenterol 2005;24:222-223]

Signet cell carcinoma of the ampulla of Vater is not common. Gardener et al. were the first to report this histological pattern in 1990. Since then, seven cases have been reported till date. However the previously reported cases were aged 49-83 years.

A 32-year-old lady presented with subacute intestinal obstruction. Exploratory laparotomy revealed adherent loops of ileum; the mesenteric nodes were enlarged. Histology of these lymph nodes showed reactive hyperplasia. On the tenth postoperative day, the patient developed jaundice, with total bilirubin 2.7 mg/dL (conjugated 2.0). Serum alkaline phosphatase was also raised (293 U/L); serum transaminases were within normal limits. Endoscopy showed normal esophagus, stomach and first part of duodenum. However, an ulcer was seen in the second part, with stricture. Biopsy from the lesion revealed fibrino-purulent exudates with clumps of highly suspicious cells entrapped in it. CT scan revealed dilated common bile duct with deformed duodenum. Multiple space-occupying lesions suggestive of metastasis were seen in the liver. CT-guided fine needle aspiration cytology
from liver revealed metastatic epithelial tumor. Endoscopic biopsy was again taken from the periampullary region; histology of this biopsy revealed signet ring cell adenocarcinoma (Fig).

The patient refused further intervention and was lost to follow up.

A review of 26 resected ampullary carcinomas by Talbot et al in 1988 revealed intestinal adenocarcinoma in 25 with one case having papillary carcinoma. Signet ring cell carcinoma of the ampulla of Vater is very rare. Our patient is the youngest reported so far. Fatigue, post prandial abdominal pain, jaundice, fever and nausea were the presenting symptoms in the previously reported cases. None of the patients had evidence of metastatic disease, except one who had spread of the disease to thoracic vertebra. Our patient had initial presentation with pain in abdomen and vomiting with abdominal distension, and was provisionally diagnosed to have subacute intestinal obstruction. We also found metastatic lesions in the liver, which has not been described previously.

Experience from previously reported cases suggests good prognosis till the first postoperative year, provided there is no metastasis at the time of surgery. Whipple’s resection, local transduodenal excision and pylorus-preserving pancreatoduodenectomy have been done with good results in these patients. Metastasis suggests poor prognosis. However, chemotherapy with 5-fluorouracil and leucovorin has been reported to be effective in increasing the survival time with good quality of life.

References

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