



A RARE CASE REPORT OF CHOLANGIOCARCINOMA PRESENTING AS MANDIBULAR SWELLING

Surgery

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ABSTRACT

Cholangiocarcinoma is difficult to diagnose and has a poor prognosis with five year survival rates of <5% due to insidious progression. Here we report a rare case of T4N0M1, Stage IVB cholangiocarcinoma presenting as a mandibular swelling. The disease had rapid progression after presentation so palliative treatment plan was undertaken with ERCP and stenting and jaundice was relieved. Here we present a rare case report of metastatic cholangiocarcinoma, its unusual presentation, investigations done to reach the diagnosis and palliative treatment given.

KEYWORDS

Cholangiocarcinoma, mandibular mass, ERCP, Stenting, CA19-9, kltaskin.

Introduction.

Cholangiocarcinoma is a malignancy that arises from cholangiocytes, which are epithelial cells that line the bile ducts. It is the second most common primary liver cancer after hepato cellular carcinoma, and is rare in many parts of the world, including Europe and the USA, where it accounts for <1% of all malignant tumors.

Cholangio carcinoma is difficult to diagnose and has a poor prognosis with five-year survival rates of <5%, due to its tendency to exhibit insidious progression.[1-3]. The oral cavity is very unusual site of metastasis accounting for only 1-2% of oral malignancy. The jawbones, particularly the mandible, were more commonly affected than the soft tissues of oral cavity.

The most common primary sites able to metastasize to the oral cavity are lung, kidney, breast, and hepato cellular carcinoma .[4-7].

The exact aetiology of CCA is unknown. There are several well-defined risk factors however, including primary sclerosing cholangitis ,liver fluke infestation, congenital fibropolycystic liver disease and intra-hepatic biliary stones.[8-11]

Other risk factors include exposure to dioxin, Thorotrast or nitrosamine. As there are no specific symptoms in early malignant lesions, patients with CCA mostly present in the advanced stages of the disease, which contributes to its poor prognosis. Oral metastases are usually evidence of a widespread disease, as occurred in this case, and indicate a grave prognosis[12]. However, in nearly 30% of patients with oral metastases, the metastatic lesion in the oral region is the first indication of undiscovered malignancy at a distant site [5]. Here we report a rare case of cholangio carcinoma presenting as painless mandibular swelling.



Figure 1: Icterus On Presentation



Figure 2: Mandibular Mass on Presentation

CASE REPORT A 55 year old female presented to OPD of Oro-maxillo-facial surgery with complaints of swelling over the left mandible since 2 months which was painless and rapidly growing. On examination a non healing ulcer over the left pterygo-mandibular raphe was seen for which an edge biopsy was done. On follow up the swelling rapidly increased in size over next 10 days and patient developed rapidly progressive painless jaundice for which she was referred to Dept of gastroenterology for evaluation of jaundice.

On examination patient had a swelling over the left cheek with size of 7*4cm situated mainly over the left side of mandible mainly over the ramus. The swelling was hard in consistency and fixed to the underlying mandible. Intra orally the swelling was not visible only a non healing ulcer was seen over left pterygo-mandibular raphe. Oral hygiene was poor. The swelling was free from overlying skin. Neck examination revealed no abnormal findings or lymphadenopathy.

On examination patient had right hypochondrial surgical scar extending as midline laparotomy scar of which no reports were available. Patient underwent the surgery 20 years back.



Figure 3: Ulcer over the Pterygo mandibular raphe



Figure 4: Old surgical scar

CT NECK-showed large destructive, expansile, heterogeneously enhancing mass with soft tissue component and irregular periosteal reaction seen epicentered at the left retromolar trigone, ramus and coronoid process of the mandible on left side along with another erosive lesion in left parietal bone on left side. Features suggestive of

aggressive malignant lesion epicentred in the ramus of left mandible with local extension and left parietal metastatic lesion.



Figure 5: CT neck showing Mandibular Mass

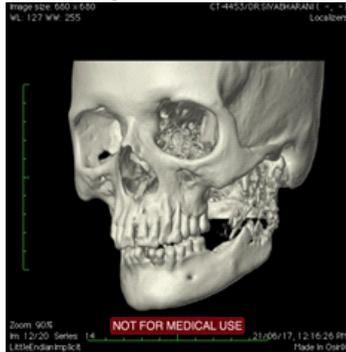


Figure 6: 3D reconstruction showing mandibular destruction

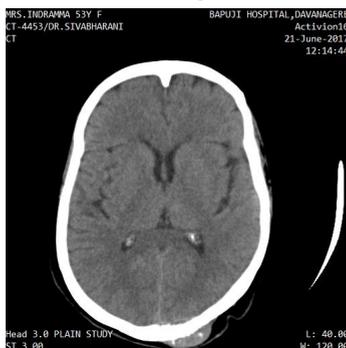


Figure 7: CT Showing Left Parietal destruction



Figure 8: MRCP showing Klatskin Tumour



Figure 9: CT showing Mandibular destruction

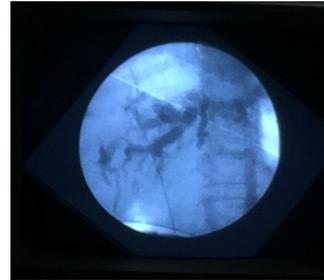


Figure 10: ERCP cholangiogram showing Klatskin Tumour

LABORATORY INVESTIGATIONS - ON ADMISSION

Total Bilirubin 11.5mg/dl
Direct Bilirubin 9.8mg/dl
SGOT 327.9 U/L
SGPT 119.1 U/L
ALP 184.6 U/L
GGT 1762.1 U/L
CA19-9 1111.9U/ML

USG ABDOMEN PELVIS-

Showed central IHBR dilatation with hyperechoic focus of 1.5cm noted in CBD with abrupt CBD narrowing.

MRCP-

Showed ill defined enhancing lesion noted at confluence of hepatic ducts with IHBR dilatation with dilated right and left hepatic ducts upto the level of confluence with abrupt cut off of common hepatic duct. CBD was not visualized. Gall bladder was not visualized ?post cholecystectomy status. Features were suggestive of klatskin tumors.

TISSUE DIAGNOSIS-

Fnac of mandibular swelling showed metastatic adenocarcinoma to the left half of mandible.

TREATMENT

In view of advanced lesion and suspected secondaries in the mandible and skull, with elevated CA19-9 levels; palliative ERCP was planned. ERCP with plastic stent insertion was done as patient was not affordable for metallic stents. Cholangiogram showed grossly dilated right and left biliary tree with abrupt cut off at hepatic confluence consistent with klatskin tumor. Post stenting the bilirubin levels came down from 11.5mg/dl to 3.8m/dl over 9 days. Patient recovered well and was symptomatically better. Four cycles of chemotherapy using gemcitabine and cisplatin were given and patient tolerated the chemotherapy well.

Patient was followed up at 3 months post chemotherapy and patient was symptomatically better with LFT within normal limits.



Figure 11: Post stenting reduced Icterus

DISCUSSION- Chemotherapy has been used in an attempt to control disease and to improve survival and quality of life in patients with un resectable, recurrent or metastatic CCA. In a retrospective study of 93 patients with un resectable or metastatic CCA, those treated with chemotherapy had a significantly higher median overall survival than those who did not receive chemotherapy (P%0.002.[14-15]

CCA has been shown to be resistant to common chemotherapy. Numerous drugs have been tested alone and in combination. The response rate has been unacceptably low. Gemcitabine has been suggested as an alternative for patients with un resectable CCA; a

survival benefit remains to be proven in a randomized controlled trial [16]

The time from the appearance of the metastasis to death is usually a few months [13].

The patient succumbed to the disease 5 months after chemotherapy.



Figure 12: Post Chemotherapy reduced mandibular swelling

CONCLUSION-

Here we present a rare case report of metastatic cholangio carcinoma which presented in the terminal stages with metastasis to the mandible and left parietal bone. The course of the disease after presentation was rapidly progressive within a span of 2 months. Unresectable end stage cholangio carcinomas can be best treated with palliation as in our case we did ERCP to relieve obstructive jaundice and we started chemotherapy as surgical resection was not possible in our case. As in our case the etiology remains idiopathic as the only relatable cause is post cholecystectomy as patient had no reports of the surgery that was done and cholangio carcinoma can't have an indolent course for 20 years.

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