Klatskin Tumor

Klatskin tumors are a type of cholangiocarcinoma. Cholangiocarcinoma is cancer of the bile duct system in the liver. It is found at the liver hilum and can also be called hilar cholangiocarcinoma. The hilum is where the left and right bile ducts join and leave the liver. These two ducts join to make one duct that drains into the bowel.

What causes a Klatskin tumor?

It is not known what causes a Klatskin tumor. There are risk factors that can cause bile duct system inflammation that could be related to Klatskin tumor development. These risk factors include primary sclerosing cholangitis, parasitic infections, ulcerative colitis, viral hepatitis, cirrhosis, bile duct stones, and choledochal cysts. Other possible risk factors include aging, alcohol use, diabetes, obesity, family history, inflammatory bowel disease, smoking, chronic pancreatitis, infection with HIV, and exposure to radioactive chemicals.

What are the symptoms of a Klatskin tumor and how are they diagnosed?

Symptoms are often caused by the tumor blocking drainage from the bile duct. These include:

- Jaundice, is the buildup of bilirubin that leads to yellowing of the skin and eyes.
- Clay-colored stools are caused by the lack of bile in the bowel. Bile normally darkens the stool.
- Bilirubinuria which is dark-colored urine.
- Pruritus (itchy skin).
- Weight loss.
- Abdominal pain.
- Enlarged lymph nodes.
- Increased liver size.
- Fatigue.

Jaundice is the most common symptom of cholangiocarcinomas. If you are jaundiced or having nonstop or worsening abdominal pain, then imaging of your abdomen should be done to find out the cause. Lab tests are also done to check the liver function tests (AST, ALT, and total bilirubin). If a mass or tumor can be found on a CT scan, MRI, or ultrasound, a biopsy should be done to confirm the diagnosis. Endoscopic retrograde cholangiopancreatography (ERCP) is a procedure commonly used to take a biopsy of a bile duct mass, and it allows for the placement of a stent to open the duct and relieve jaundice.

How is a Klatskin tumor treated?

Treatment options include surgery, liver-directed therapy, radiation with or without chemotherapy, chemotherapy, and targeted therapy. Klatskin tumors can involve the liver and one or both bile ducts. If the tumor only involves one side of the liver and/or one side of the bile duct, it may be possible to surgically remove it.

If surgery is not possible, radiation therapy can be used with or without low-dose chemotherapy. Low-dose chemotherapy can make the tissue more sensitive to the radiation. Liver-directed therapy such as

chemoembolization or radioembolization can also be administered if confined to one lobe of the liver but is not able to be surgically removed.

Chemotherapy can be used with radiation or as the main treatment when the tumor is not able to be removed. Chemotherapy medications that are used include: fluorouracil, gemcitabine, cisplatin, capecitabine, oxaliplatin, albumin-bound paclitaxel, and liposomal irinotecan.

Targeted therapies can be used based on molecular testing and they include:

- NTRK gene fusion-positive tumors: entrectinib and larotrectinib.
- MSI high or dMMR tumors: pembrolizumab and dostarlimab-gxly.
- BRAF-V600 mutated tumors: dabrafenib with trametinib.
- FGFR 2 fusions or rearrangements: pemigatinib and infigratinib.
- IDHI mutations: ivosidenib.
- Tumor Mutation Burden -High (TMB-H) tumors: nivolumab with ipilimumab, pembrolizumab.
- RET gene fusion positive: pralsetinib, selpercatinib.
- HER 2 positive: trastuzumab and pertuzumab.

Patients who have not previously been treated with immunotherapy may be prescribed nivolumab, lenvatinib, and pembrolizumab or durvalumab. These agents can be used as single agents or together with chemotherapy or other targeted therapies. Other targeted therapies that can be used include regorafenib.

Treatment is specific to your case. Be sure to talk to your provider about any questions or concerns you have. Clinical trial options may also be available.