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Neuroendocrine Tumor: A Rare Presentation

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Abstract:
Surgical OPD have variety of patients. Amongst these there are intra-abdominal lumps, which may be painful or painless and can be symptomatic or asymptomatic and are investigated on their merits. A person with an abdominal mass may suffer an increase in weight and symptoms such as abdominal discomfort, pain, and bloating
Abdominal masses are mostly treatable. However, complications may arise depending on the cause of the mass. Abdominal masses can be the result of a number of factors, like an injury, cyst, benign tumor, cancer, or other diseases.

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I. Introduction

A neuroendocrinal tumor begins in endocrinal cells as well as nerve cells. These cells are found at many places like in lungs, gastrointestinal tract, pancreas, thyroid, etc. and maintain the mechanics like food transit time, rate of air and blood flow. There are many types of neuroendocrinal tumors but chiefly they are:

- Pheochromocytoma: Pheochromocytoma is a tumor that begins in the chromaffin cells of the adrenal medulla. These specialized cells release stress hormones. . This type of tumor increases the production of the hormones adrenaline and nor-adrenaline, which affects blood pressure and heart rate. These tumors may be benign, but still life-threatening as they release large amounts of stress hormones into the bloodstream after injury, affecting haemodynamic stability of the patient. 80% of these tumors are unilateral and only about 10% are bilateral and very few patients may have these tumors outside adrenal gland.
- Merkel cell cancer: Merkel cell cancer, or trabecular cancer of skin, is a malignant tumor of merkel cells, mostly affecting elderly. It usually begins in hormone-producing cells of skin and hair follicles. It is four times more common in women than in men highly aggressive, or fast-growing, rare cancer.
- Neuroendocrine carcinoma: Nearly more than half of neuroendocrine tumors are nothing but neuroendocrinal carcinoma. Neuroendocrine carcinoma can be found in various organs, like the lungs, brain, and gastrointestinal tract.

People with a neuroendocrine tumor may experience the following symptoms or signs. Sometimes, people with a neuroendocrine tumor do not have any of these changes. Or, the cause of a symptom may be another medical condition that is not cancer. These tumors may have the following presentation:

- Hyperglycemia
- Hypoglycemia
- Diarrhea
- Pain
- Lump
- Jaundice
- Weight loss
- Weight gain
- Unusual bleeding or discharge
- Unusual bleeding or discharge
- Persistent fever
- Acid peptic disease
- Skin diseases

Diagnosis is usually done by taking biopsies but may not be possible in many cases. Then other tests are done to aid in the diagnosis of neuroendocrine tumor, which may be:

- Routine blood tests and sugar levels.
- Urine examination, particularly for excretion of metabolites of adrenaline and nor-adrenaline{5-HIAA}
- CT scan

Evaluation is done not only for diagnosis but to stage the disease also. The various stages of neuroendocrine tumor can be as follows:

- Stage 1: Primary tumor less than 2 cm
- Stage 2A AND 2B: Tumor more then 2cm size but not spreading to nearby structures and lymph nodes
- Stage 2C: Tumor spreading to nearby structures like bone, muscles or cartilage but not to lymph nodes
- Stage 3: Tumor spreading to nearby lymph nodes
- Stage 4: Metastasis

Treatment options

Treatment will depend on the type of tumor, staging, possible side effects, etc.
Surgery is the main treatment option particularly for phaeochromocytoma and merkel cell cancer.
Radiotherapy is advocated for tumors which have metastasized or are difficult to operate.
Targeted therapy aims at modifying the specific genes, proteins, tissue environment which are important for tumor growth. Hence, it blocks the growth and spread of tumor cells.
Interferons are being used but its effectiveness is uncertain.
Regular follow up of patient is required for years ahead as recurrence can be immediate or late. [3][4]

Case Presentation:

A 66 year old male patient presented with large intra-abdominal lump occupying the whole of abdomen since years. Lump was asymptomatic and had no pressure symptoms. Patient had been previously investigated at many other centers. General examination and all routine blood investigations were normal.

CT abdomen showed a large intra-abdominal lump possibly teratoma with no enlarged lymph nodes.

Laprotomy was planned and exploration of the abdomen done. A large intra abdominal lump found which appeared to arise from root of mesentry. No lymph node enlargement was seen. All viscera found to be normal. Lump was removed along the involved bowel was resected and anastomosed. Post operatively patient expired due to sudden cardio respiratory arrest. Follow up of the patient showed a progressive recovery. Histopathology examination of the tumor was suggestive of neuroendocrine carcinoma.

II. Conclusion

A neuroendocrine tumor is very rare type of tumor with varied presentation. It has multi treatment options, but with variable prognosis. As this tumor has variety of presentation, its diagnosis is difficult and requires a lot of clinical judgment. As late recurrence of the tumor is possible, a long term follow up is required. This tumor shows predominance in white skinned people and in people who are generally above 50 years of age. In India approximately 1 million cases are diagnosed annually. The five year survival rate of this tumor is nearly 60%. The prognosis is much better if tumor is discovered early and has not spread to lymph nodes or has not metastasized.

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