Primary Squamous Cell Carcinoma of Spleen- The Histological Surprise

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Abstract – The incidence of tumor in the spleen is very less. Even if they do occur, they are more commonly benign and if malignant, either lymphoma or angiosarcoma are seen. To the best of our knowledge, primary squamous cell carcinoma of spleen is very rare and has not been reported in India until now. This is the first case we are reporting from India. Mostly tissue diagnosis of primary splenic carcinomas is done after splenectomy. Our patient of elderly age group has undergone surgery as a definitive management for primary squamous cell carcinoma of the spleen. The present study provides insight into the response to standard treatment and the prognosis of the primary splenic squamous cell carcinoma for a single case.

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INTRODUCTION

I. Case Report

The primary epithelial tumors of spleen is extremely rare. Most of the time these kind of tumors are known as "Histological Surprises". These surprises we came to know either after splenectomy or in autopsy. The incidence of splenic secondary tumors at autopsy was 0. 6% and at splenectomy, 1.1%.(2). As no other source of primary epithelial tumor metastasis can be identified , we have to mark in this tumor, spleen as primary origin as primary splenic squamous cell carcinoma. To the best of our knowledge, only one case has been reported of primary squamous cell carcinoma in China(1) and one case of metastatic squamous cell carcinoma until now. However, primary squamous cell carcinoma of the spleen with no identified lesion in other organs is very rare. The study aims to demonstrate the treatment outcomes in the elderly age group and overall survival after definitive surgery.

Case presentation – An averagely built, 82 year Hindu gentleman hailing from a small village of north India was admitted to Sawai Man Singh Hospital, Jaipur, Rajasthan, India, on 05 July 2021 with complaints of left hypochondriac swelling for the last 30 years and insidious onset of dull aching pain for the last 6 months. Patient is a chronic tobacco chewer from last 6 year, with no other co morbidity. On physical examination, there was a large solid hard mass arising from the left hypochondriac region and extending towards the umblical region which was moving with respiration and was of size approximately 12 x 10cm felt below the left subcostal margin, the upper border could not be appreciated due to merging of the upper margin of the mass below the left costal margin and the fingers could not be insinuated between the ribs and the mass. Mass was non-tender with smooth surface & hard consistency. On routine blood investigations, the Hb. was 12.8gm/dL, TLC - 11k /cu mm, and TPC-1.76 lakhs/cu mm. The other biochemical and organ function tests suggested no abnormality. On Radiological examination, contrast enhanced computed tomography (CECT) showed an enlarged spleen measuring 17 cm with peripheral enhancement and a centrally necrotic fluid density lesion measuring approx. 16x 9 cm with the lower pole parenchyma. Curvilinear calcifications were also seen in the wall of this lesion. Differential diagnosis of Benign Splenic tumor & calcified hydatid cyst was considered. Subsequently, an exploratory laparotomy was done which revealed a hard splenic mass for which the patient underwent splenectomy.

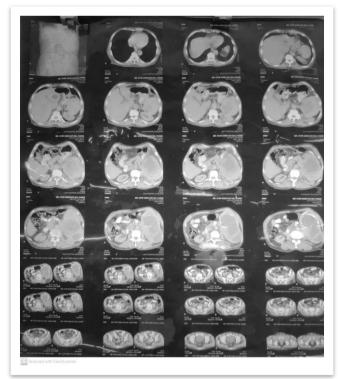


Fig.1 CECT Whole Abdomen showing enlarged spleen of size 17cm with peripheral enhancement & central necrosis.

Post Operative Pathological Evaluation - The pathological analysis revealed a gross specimen of a large splenic mass of approximately 18 x 12 x 12 cm on the cut section, large cystic areas with thick septations were seen. On microscopy, tumor nest cells surrounded with desmoplasia were seen and the peripheral capsular margin showed focal areas of invasion. Histopathological features of the section were suggestive of moderately differentiated squamous cell carcinoma.



Fig.2- Gross Morphology specimen of spleen in cut section shows Cystic lesion due to central necrosis.

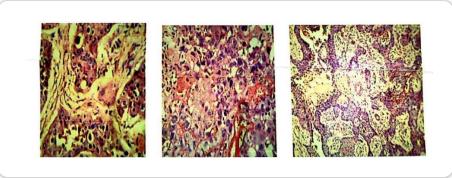


Fig.3- Histomorphological features showing nests of tumor cells Surrounded by desmoplastic cells. Focal area of invasion.

Follow – Since the patient was denied for further chemotherapy or radiotherapy, he was discharged in good medical condition on 15/07/2021. Further follow-up investigations were completely within normal limits. Patient is currently alive and healthy, which suggests a good prognosis of the disease after definitive surgery.

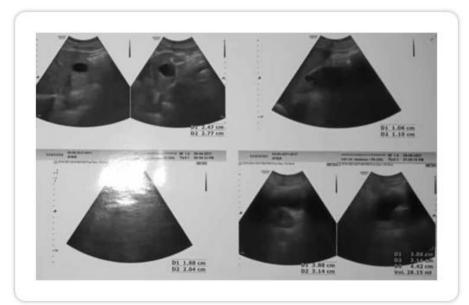


Fig.4- Post Operative Sonography done after 5 months



Reveals no other mass abnormality. Fig.5- Post operative Scar Mark of Left subcostal incision.

Discussion

II.

As previously mentioned, this is a rare case. To Our knowledge, in the whole world only 1 case has been reported from China in 2018 and In India it seems to be the very first case, without any knowledge about the pathology. Most of the cases that have been reported are mostly metastatic tumors from other primary sources. And in our case, there is no other pathology found that seems to metastasize in the spleen.

Like every other organ, the spleen consists of stroma and parenchyma. The stroma of the spleen is composed mainly of a network of reticular connective tissue. This mesh provides support for blood cells and cells of the immune system. The capsule of the spleen consists of dense irregular fibroelastic tissue. The connective tissue of the capsule contains contractile cells called myofibroblasts. Although on admission CECT whole abdomen was done but PET CT was not done and not even tumor marker. As the results were unexpected. The actual mechanism was still not able to understood. Some literature suggest mechanism similar to endometriosis. One Mechanism may be due to trauma, which causes infiltration of the epithelial cells inside parenchyma, then over time due to inflammation & under influence of hormone there may be some mitotic changes causing squamous cell carcinoma. Now what if there is no history of trauma, then another mechanism proposed may be due to abundant blood supply of spleen, some of the epithelial cells may have migrated inside spleen, then over time under influence of hormones and inflammation, dysplastic changes have led to cause squamous cell carcinoma(3). As this tumor is extremely rare, the literature is not available and anything cannot be said with surety. Therefore, the origin and mechanism of the tumor in the present case study remains unknown. The genetic marker study for primary squamous cell carcinoma may be done which includes P63 & P40.We did not go in this process and patient also denied for further intervention. However, our patient has been in follow-up and has not complained any other problem.

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