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Pure Testicular Teratoma Presenting as a Metastatic Germ Cell Tumor

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Abstract- Mature teratoma comprises 5-10 % of all testicular neoplasms. Pure testicular teratoma is rare in adulthood with an incidence of 5 %. Regardless of differentiation postpubertal testicular teratoma have aggressive behavior and microscopic appearance of metastasis may differ from that of the primary. We present a case of pure teratoma of testis in an adult presenting as a metastatic germ cell tumor.

Index Terms- Pure teratoma, Testis

I. INTRODUCTION

Teratoma is a tumor containing tissues representative of more than one germ cell layer. Teratoma may be classified according to the degree of maturation or differentiation of these tissues into mature and immature. Mature teratoma consists exclusively of well differentiated tissues . Immature teratoma consists of less differentiated or primitive appearing tissues occasionally with malignant transformation. Pure teratoma of the testis is a relatively rare tumor that can occur in children and adults. Teratoma occurring in the postpubertal age group is considered to be a malignant tumor with the capacity for metastasis.[1]

II. CASE REPORT

A 24 year old boy presented with retroperitoneal mass of 2 months duration. On exploratory laprotomy 6 x 4 cm right iliac lymph node mass was found . Excision biopsy of the mass was done. Histopathologic examination of the lymph nodes showed malignant cells with large vesicular nuclei, prominent nucleoli arranged in nests, trabeculae and tubular pattern. Also seen were foci of microcystic pattern and Schiller -Duval bodies.

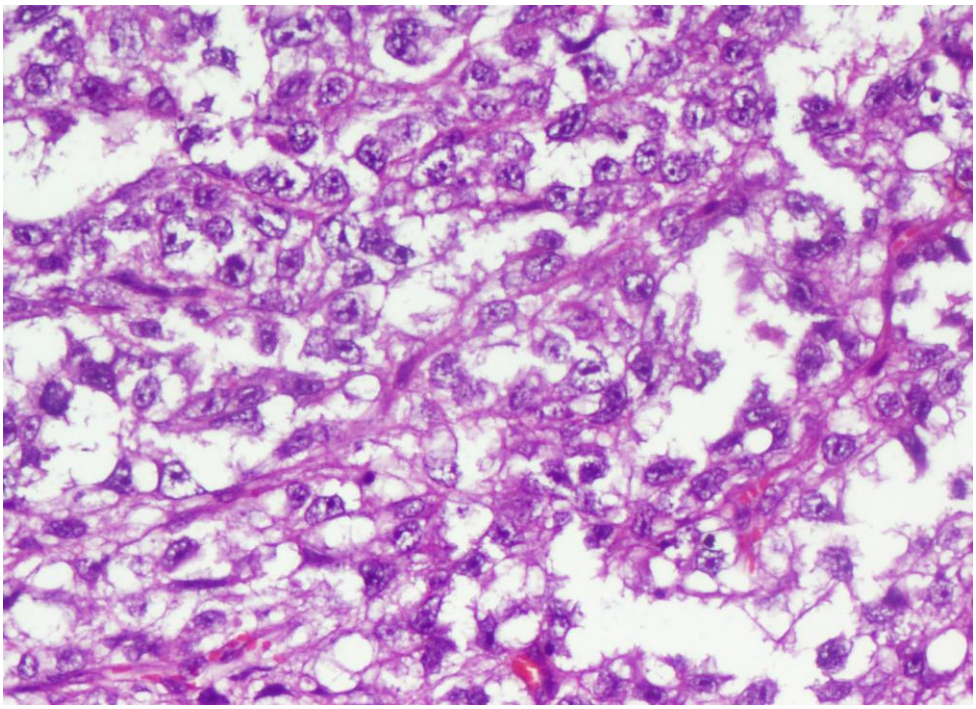

Fig 1.Retroperitoneal node showing malignant germ cell in nests and trabeculae H&E x 200

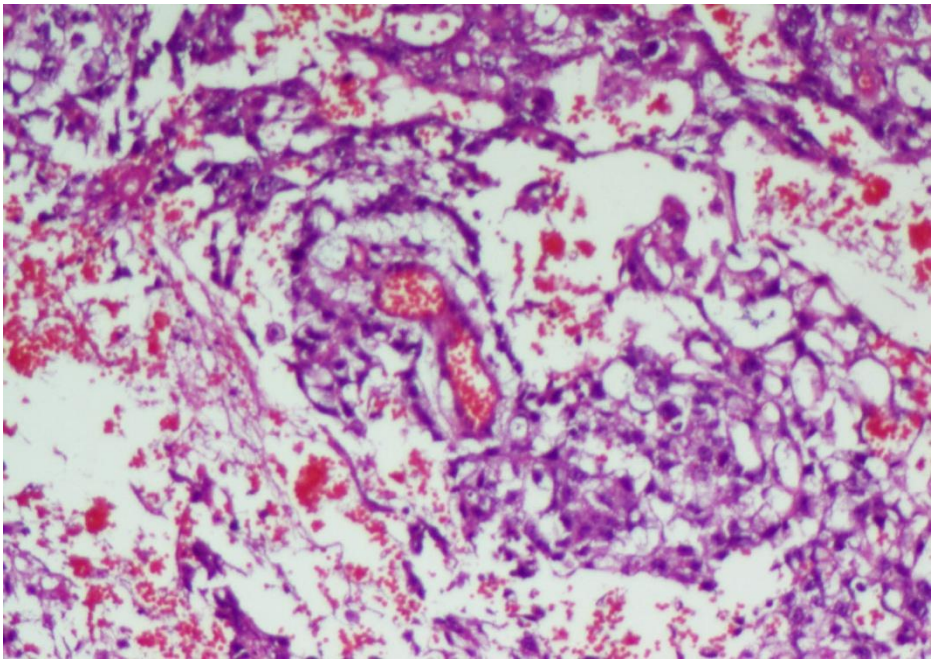

Fig 2. Schiller-Duval bodies H&E x200

A diagnosis of metastatic mixed germ cell tumor with embryonal carcinoma and yolk sac tumor components was given . The patient on further investigation for a possible primary revealed a small nodule in the testis and elevated serum β HCG & alpha-fetoprotein levels. Patient underwent a high inguinal orchidectomy. The specimen of testis was normal in size. Cut section of the testis showed a small white nodular lesion 2 x 1.5 cm in size.

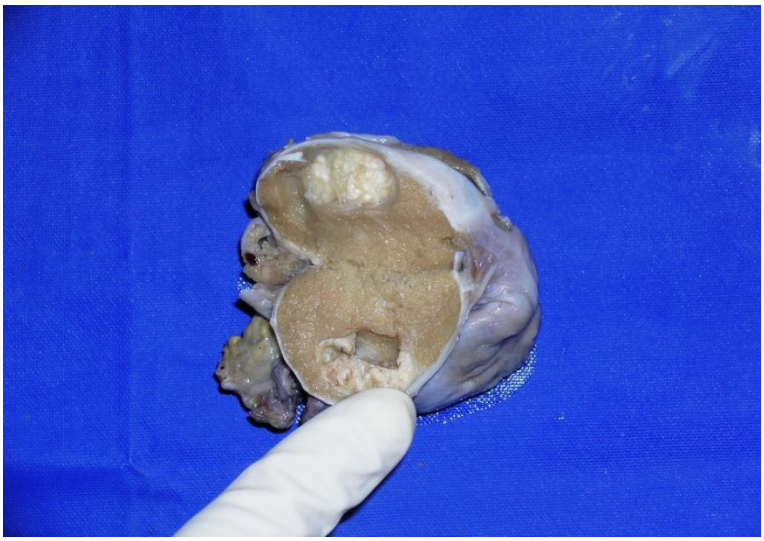

Fig 3 .Testis normal in size with a grey white nodular lesion.

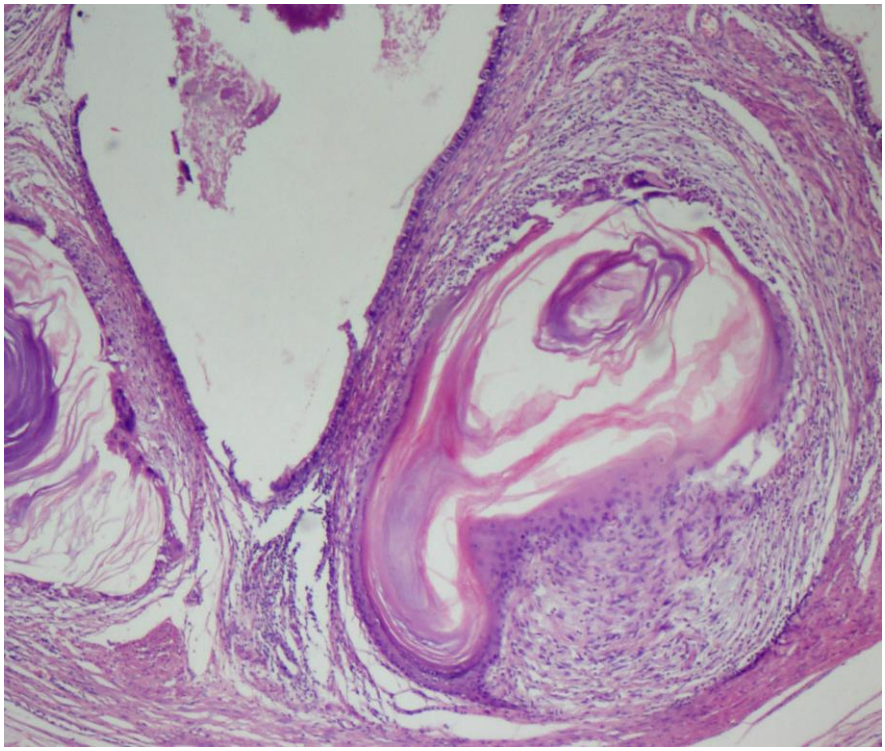

Fig 4. Testis showing a mature teratoma H&E x 100

Histopathologic examination showed mature teratoma .Multiple sections studied from the tumor in the testis showed no immature elements or evidence of other germ cell tumor components .

III. DISCUSSION

Testicular tumors can be classified into germ cell tumors and non germ cell tumors. Approximately 95 % of testicular tumors arise from germ cells .Germ cell tumors are divided into seminomas which occur in approximately 40% of the population and non-seminomatous tumors (NSGCT) which may be seen in pure or mixed form. [2,3] NSGCTs are histopathologically heterogenous and commonly consists of embryonal carcinoma, yolk sac tumor, teratoma and choriocarcinoma .

Teratoma is a tumor containing tissue elements derived from different germ cell layers (endoderm, mesoderm, and ectoderm). While teratomatous elements are found in 55-85 % of all NSGCTs in adults, only 2-6 % of testicular teratomas are pure teratomas.[3,4 ,5] Pure teratomas can be further subdivided by the sexual development of the patient into pre pubertal and post pubertal testicular teratomas. Their biology appear to be significantly different. Prepubertal teratomas usually follow a benign course, successfully managed with orchidectomy, do not recur and do not metastasize. Postpubertal teratomas are regarded as malignant, capable of metastatic behaviour, irrespective of whether the elements are mature or immature. Metastasis may be teratomatous and/or consist of other nonseminomatous germ cell elements, such as embryonal carcinoma and choriocarcinoma.[1,3]

Teratomas are the second most common germ cell tumors after yolk sac tumors in the prepubertal age group . They occur most commonly as pure teratomas in this age group The mean age of patients at diagnosis is 20 months, and rarely occur after the age of 4 years. Postpubertal teratomas are usually found in association with other germ cell elements (mixed germ cell tumors) and occur mainly in the second to fourth decades of life or sporadically in those older than 50 years.

Mixed germ cell tumors contain more than one germ cell component and are much more common in the testis than any of the pure histologic forms, representing 32%-60% of all germ cell tumors. Most common admixtures being embryonal carcinoma and teratoma . Minor foci of yolk sac tumor are common, although it is usually overshadowed by other components, such as embryonal carcinoma.

The biologic behavior of teratomas is quite variable, depending on the pubertal status of the testis. In prepubertal testis, pure teratomas are considered benign even when they are histologically immature. This benign behavior has led some investigators to recommend a testis-sparing tumor enucleation rather than orchidectomy. However, such conservative treatment is not an option for teratomas in postpubertal testis. Of important distinction, every element in a postpubertal testicular teratoma (mature or immature) can metastasize, irrespective of its histologic characteristics.The metastatic disease in a pure teratoma may contain other subtypes of non seminomatous germ cell tumors in addition to teratoma.[6,7]

The exact mechanism of metastasis in a pure teratoma remains controversial .The finding of IGCNU and associated scars support the theory that undifferentiated stem cells within the primary testicular tumor metastasize and differentiate into adult teratoma or undifferentiated germ cell tumor at the new

site. The primary germ cell tumor undergoes regression and /or differentiation to mature teratoma . It, therefore, represents a differentiated, malignant neoplasm that forms through a process of "maturation" from more primitive types of germ cell tumors .[7] Malignant transformation of mature teratoma into other germ cell elements in the metastatic site may represent another possibility of development of metastasis.[7,8]

Pure teratomas with metastasis are thought to be mixed germ cell tumors in which the nonteratomatous component have undergone spontaneous regression. In many of these cases, patients present with distant metastases of germ cell tumor and are subsequently found to have evidence of a regressed germ cell tumor in the testis. For patients who present with a scarred nodule in the testis in the absence of metastases, one must distinguish a completely regressed teratoma, which has metastatic potential, from nonspecific scars that result from injuries or vascular lesions.

Epidermoid cyst and dermoid cyst need to be distinguished from the usual teratoma of the adult testis because they are uniformly benign, whereas postpubertal teratoma may have associated metastasis of either teratomatous or nonteratomatous germ cell tumors. Postpubertal teratoma are associated with changes in the adjoining seminiferous tubules – Intra tubular germ cell neoplasia (IGCNU), microlithiasis, tubular atrophy, sclerosis and scarred areas. IGCNU is absent in dermoid cysts and the surrounding testis also lacks the changes seen in postpubertal teratomas.[6]

IV. CONCLUSION

Primary pure teratoma of testis in post pubertal age are considered malignant with capacity for metastasis and there is a high incidence of malignant germ cell tumor being present in the metastasis.

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