CASE OF THE MONTH

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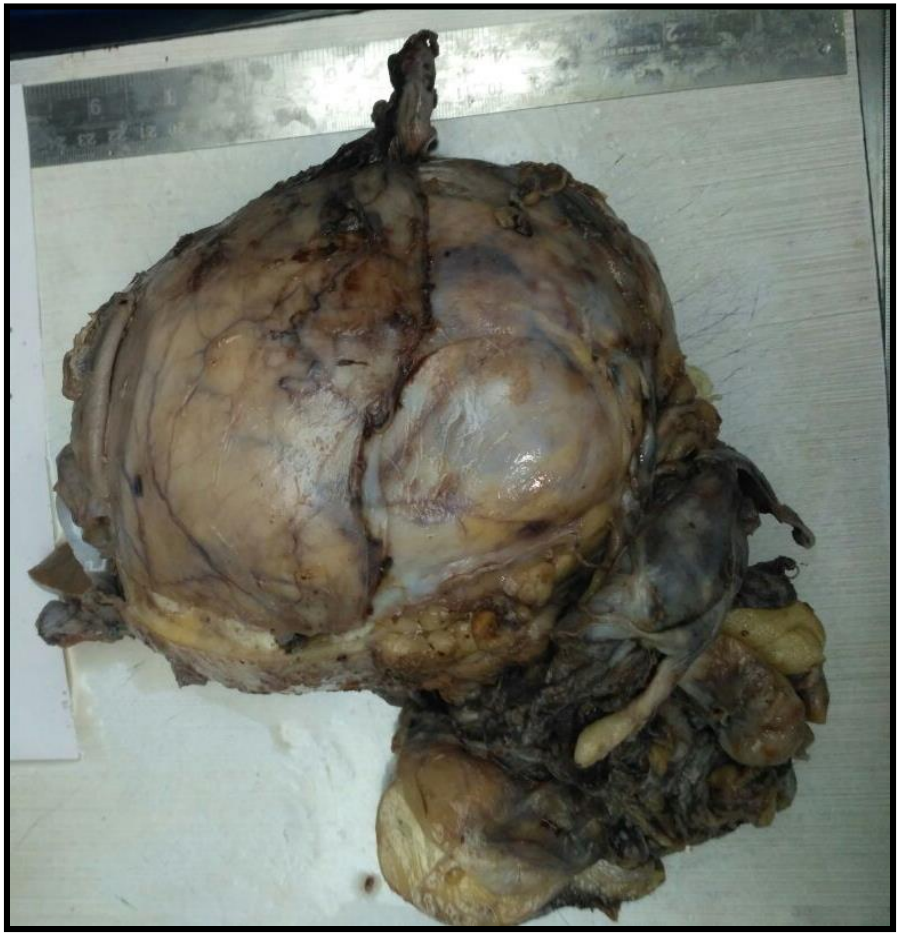
- A 50 years old male presented with a mass in the left inguinal region for duration of nine months

- After ascertaining further clinical history, radiological examination was done, which showed a non homogenous mass with variable echo intensity in the paratesticular region measuring cm in maximum dimensions. No significant lymphadenopathy seen

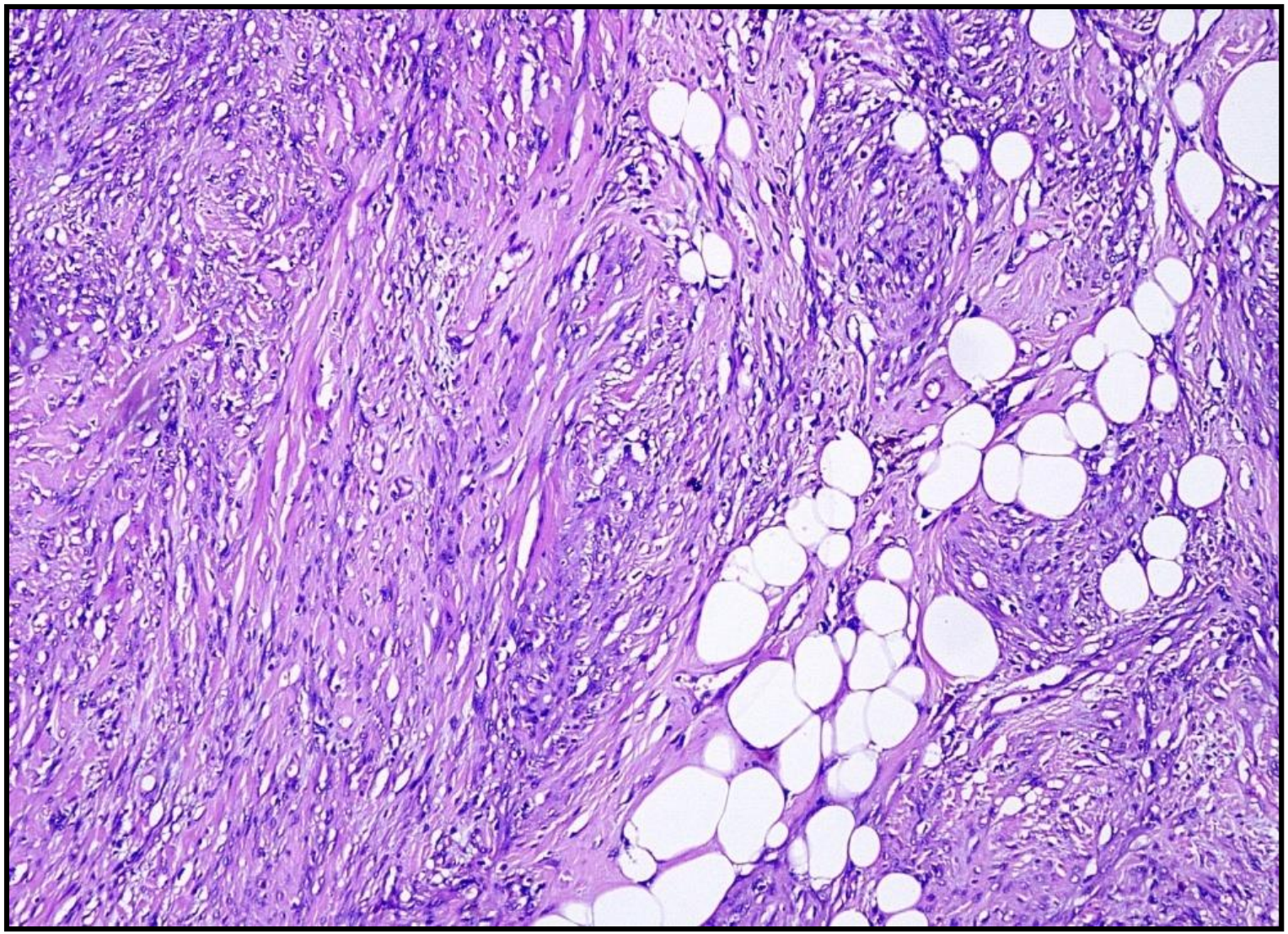
- The mass was then surgically excised and subjected to histopathological examination

GROSS APPEARANCE

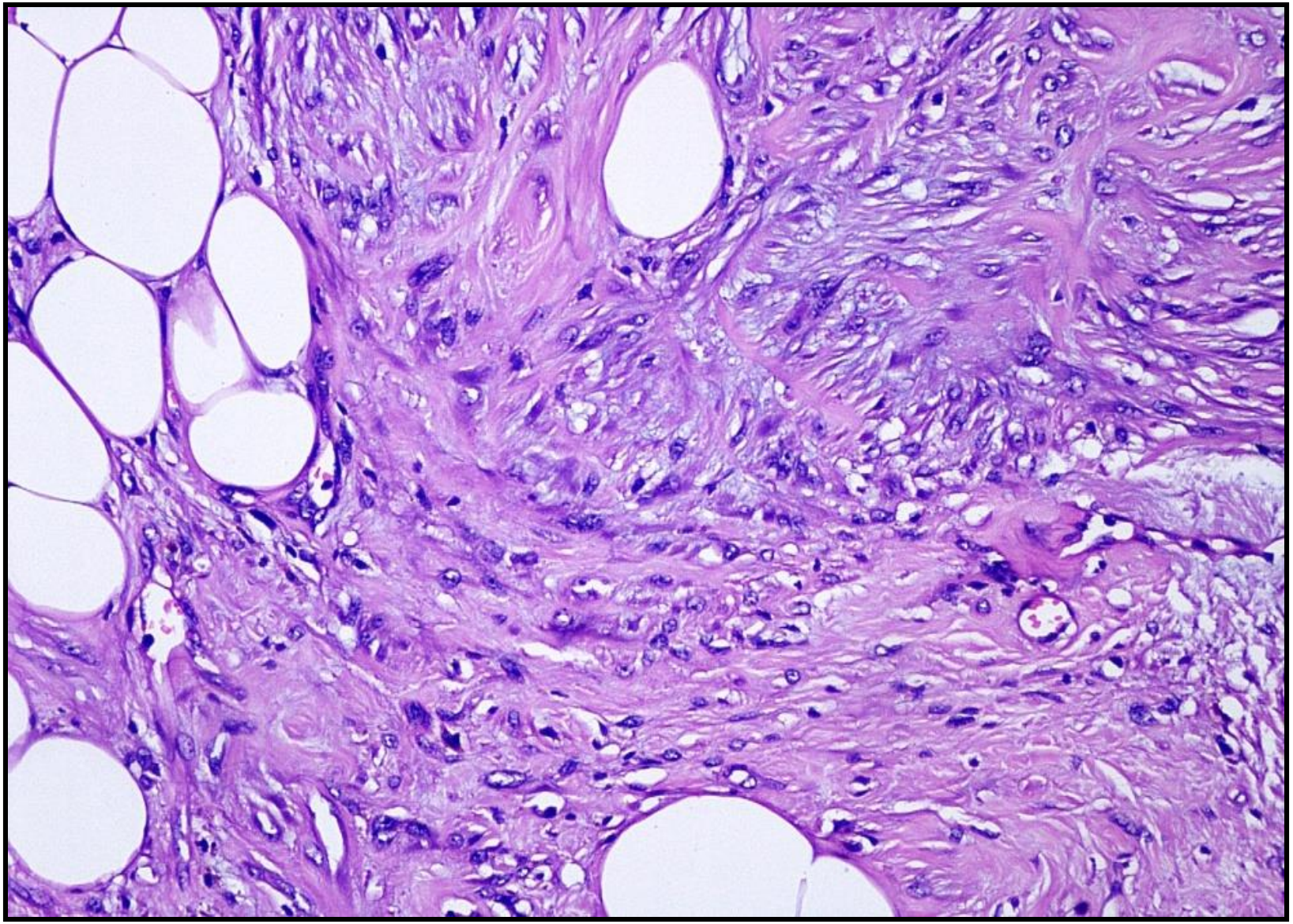


Large firm tan grey mass with lobulated cut surface, surrounded focally by fibrofatty areas

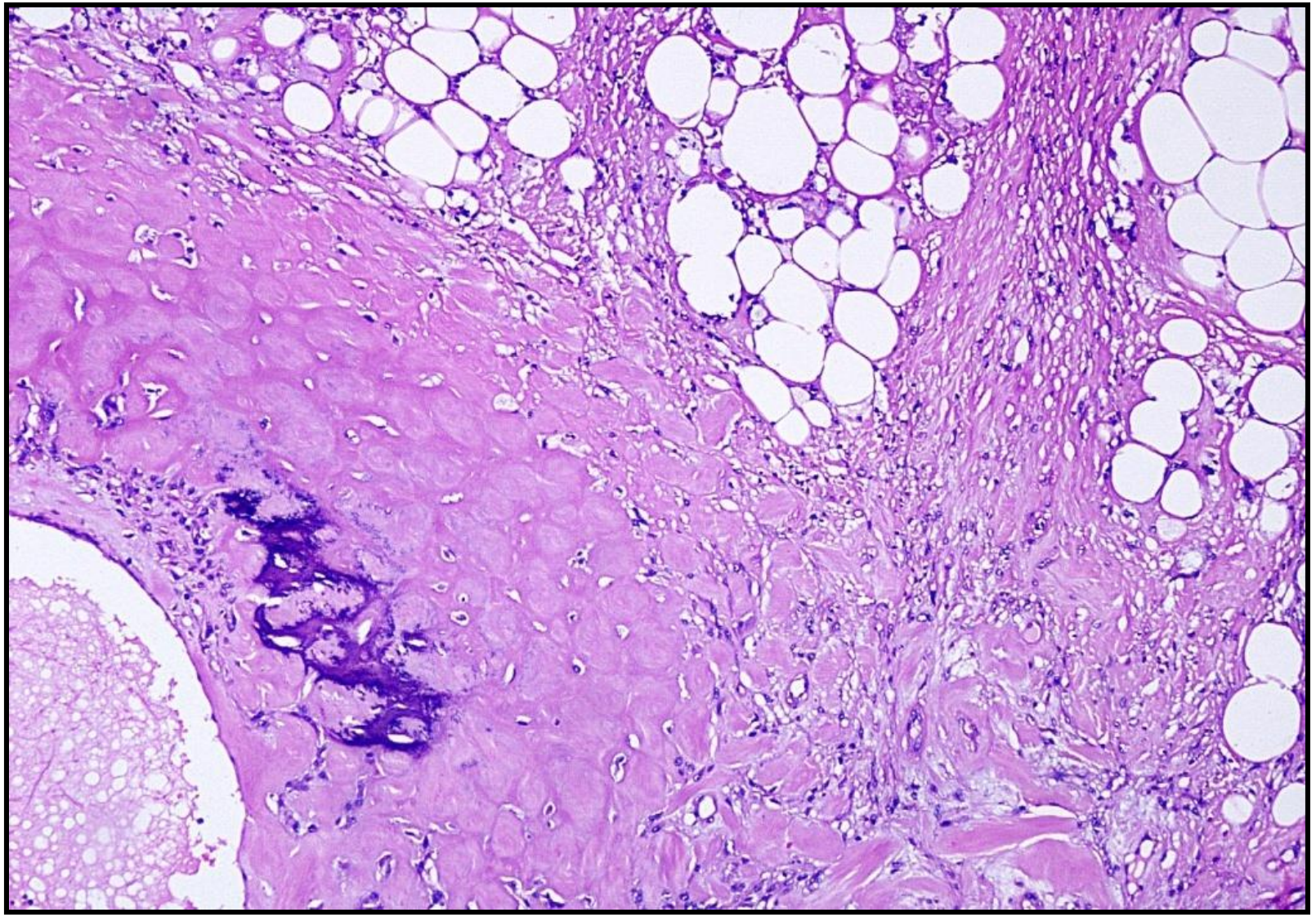
MICROSCOPY



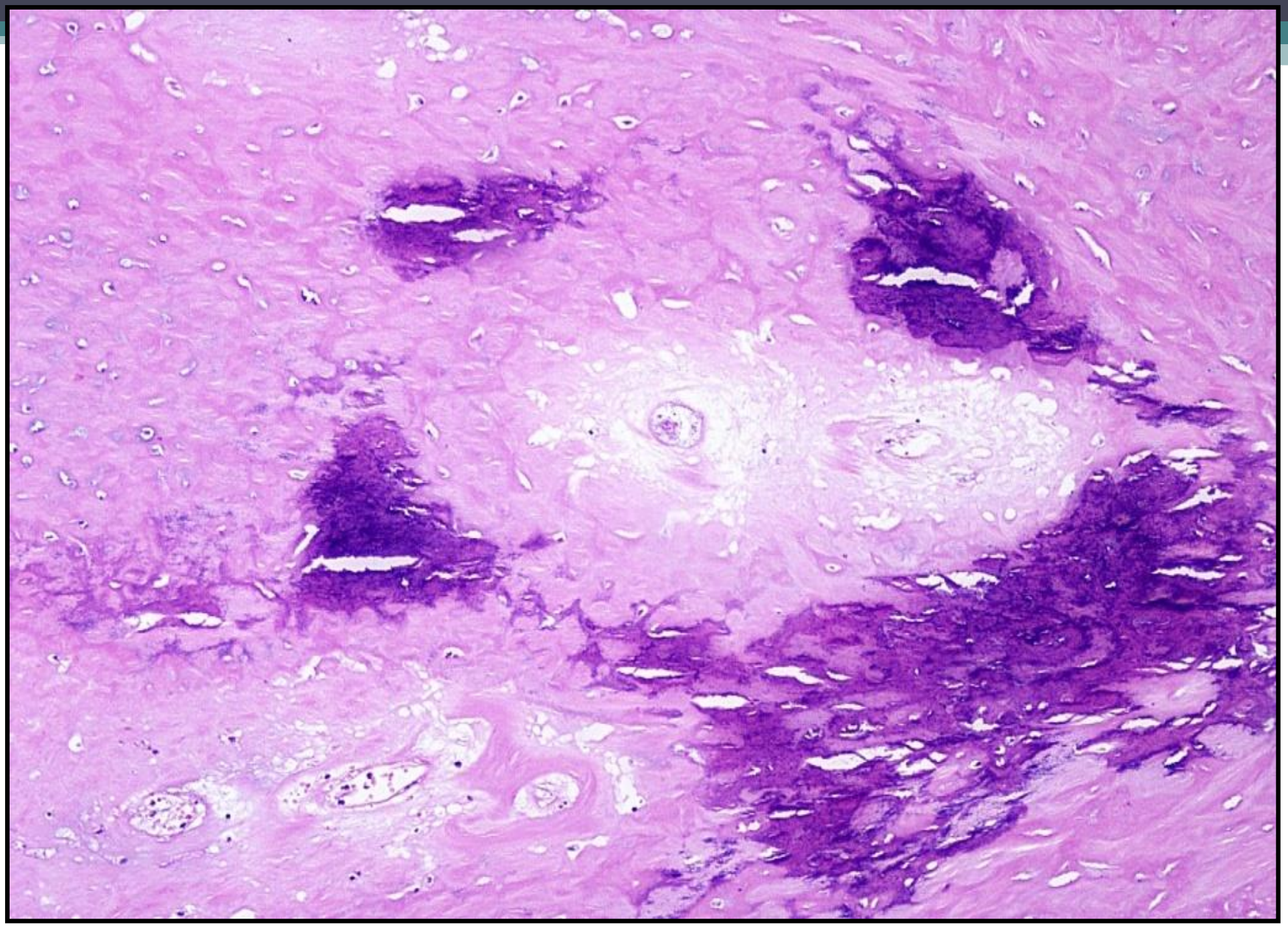
Neoplastic spindle cells in interlacing fascicles and storiform pattern, exhibiting moderate nuclear atypia, along with scattered adipocytic cells (H&E , 100x)



Neoplastic spindle cells in interlacing fascicles and storiform pattern, exhibiting moderate nuclear atypia, along with scattered adipocytic cells(H&E, 200x)



Neoplastic spindle cells in interlacing fascicles and storiform pattern, along with scattered adipocytic cells and foci of metaplastic bone formation (H&E, 100x)



Focus of metaplastic bone formation within the tumor (H&E, 200x)

Summary of histopathological findings:

- Microscopic examination revealed a relatively circumscribed tumor comprising of neoplastic spindle cells in interlacing fascicles and storiform pattern, exhibiting moderate nuclear atypia

- Scattered and clustered adipocytic cells were noted entrapped in between devoid of any significant atypia/pleomorphism.

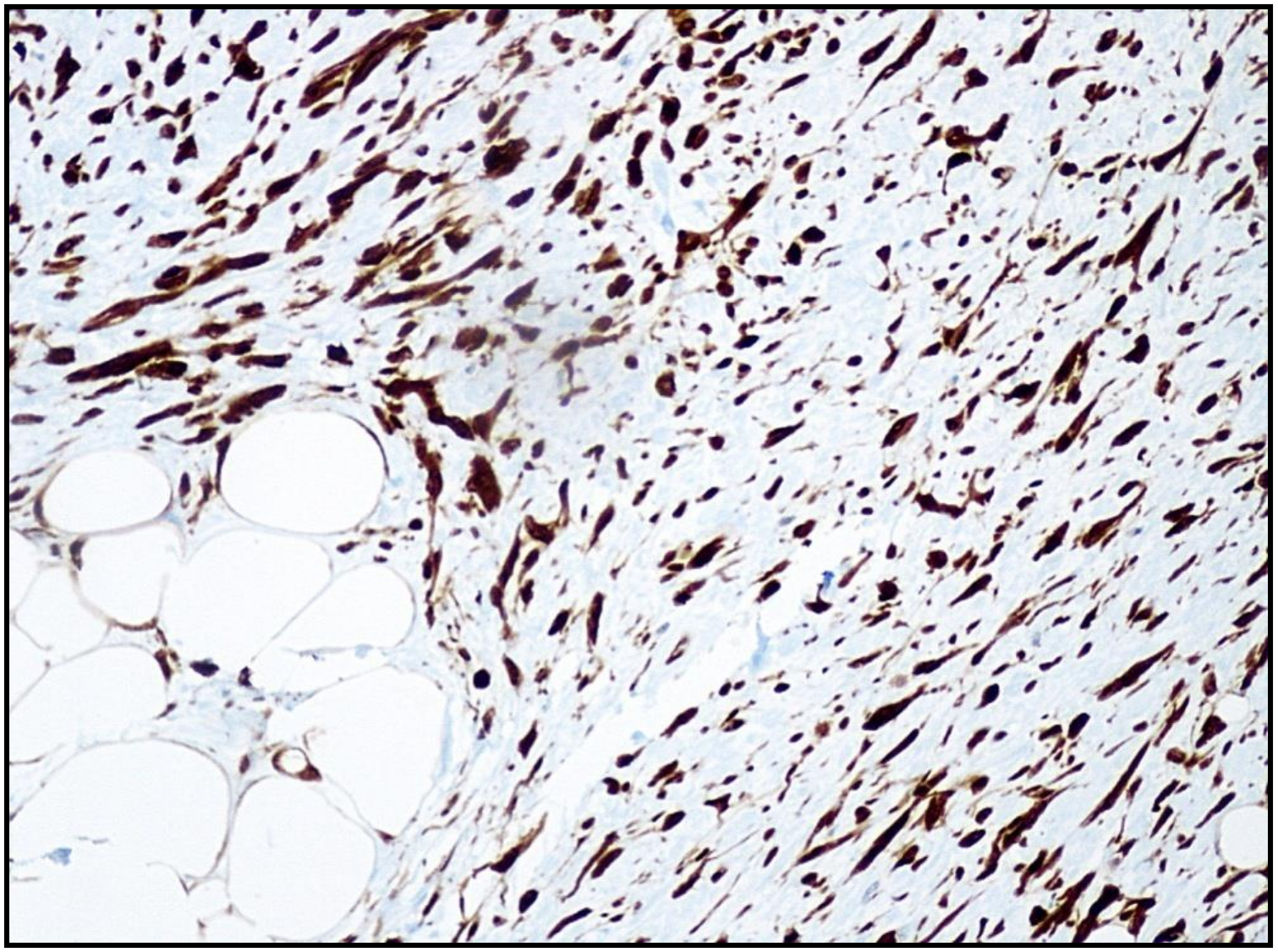
- Also noted were large foci of metaplastic bone formation in between the tumor cells

IMMUNOHISTOCHEMISTRY

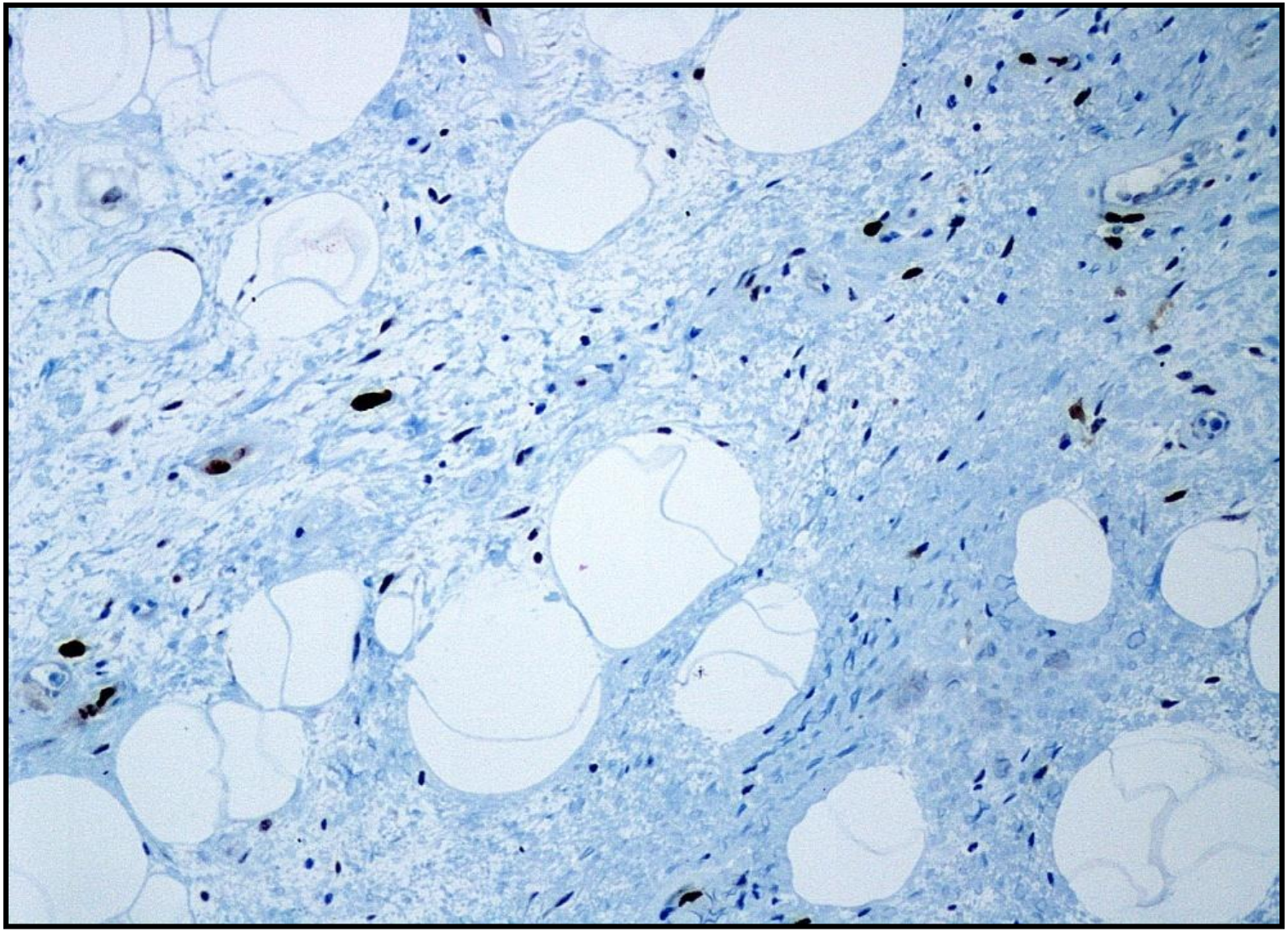
The tumor cells were positive for vimentin while negative for Ck, SMA , S100, CD34

Features were suggestive of a mesenchymal tumor of intermediate aggrressiveness.

Subsequently CDK4 and MDM2 IHC was applied



Spindle cells and lipoblasts showed diffuse and strong nuclear positivity for CDK4



30% of these cells showed nuclear expression of moderate intensity for MDM2

- The neoplastic spindle cells and well differentiated liposarcoma component showed diffuse and strong nuclear positivity for CDK4

- 30% of these cells showed moderate intensity nuclear expression of for MDM2

A diagnosis of Dedifferentiated Liposarcoma (FNCLCC grade II) arising in the background of well differentiated liposarcoma was rendered

DISCUSSION:

- This is a case report of a patient with paratesticular dedifferentiated liposarcoma with areas of osseous differentiation
- Liposarcoma is a relatively rare pathological entity, though it is a common histological subtype of genitourinary sarcomas in adults
- Liposarcoma usually occurs in the deep soft tissues of extremities and in the retroperitoneum

- Dedifferentiated liposarcoma (DDL) is a rare mixed histologic subtype defined by the association of well-differentiated liposarcoma and a nonlipogenic sarcoma of variable histological grade usually with histologically abrupt transition

- Different studies have shown that the majority of dedifferentiated liposarcomas present as de novo lesions, whereas the remainder develop as a late complication of a preexisting well-differentiated liposarcoma

- The dedifferentiated component may already be present at the time of original excision but is much more commonly seen in recurrent or metastatic foci

- In different studies, however, the majority of dedifferentiation occurred in de novo lesions

- The interval between the primary appearance of the tumor and dedifferentiation varies greatly and in most cases exceeds 5 years
- The dedifferentiation is not a site-specific phenomenon but is more likely a time dependent phenomenon
- Commonest sites involved by dedifferentiated liposarcoma are retroperitoneum, extremities, trunk, and subcutis
- Paratesticular liposarcomas, overall, are rare and typically reported as an isolated case or components of larger studies of liposarcomas
- There are no metastases and the overall prognosis is good in majority of cases

- Dedifferentiation is detected in up to 10% of well differentiated liposarcoma (WDL) of any subtype and it is related to poorer prognosis

- Histopathologically, there are two grades (low and high) of DDL

- Low grade dedifferentiation is characterized most often by the presence of uniform fibroblastic spindle cells with mild nuclear atypia

- Cytogenetics shows similar amplification of MDM2 (amplification of the 12q14 chromosome band), and CDK4, as seen with WDL

- Heterologous dedifferentiation may occur in about 5–10% of the cases (as seen in the present case) as well as osseous metaplasia, which has not been related with aggressiveness or invasiveness of the tumor

- It has been suggested that low-grade differentiation may represent a precursor lesion of high-grade differentiation

- Paratesticular well-differentiated liposarcoma can develop into either low-grade or high-grade dedifferented liposarcoma over a variable period of time

- High grade dedifferented liposarcoma (FNCLCC grade III) has been associated with a more aggressive clinical course.

- *In conclusion, though dedifferentiated liposarcoma is a rare histoloathological subtype of liopsarcoma, it should be considered as part of the differential diagnosis in tumors arising in paratesticular region*

- *Majority of dedifferentiated liposarcomas present as de novo lesions, whereas the remainder develop as a late complication of a preexisting well-differentiated liposarcoma*

- *In cases, which present as an udifferentiated mesenchymal lesion on morphology and immunohistochemistry, the IHC markers CDK4 and MDM2 have been proved as a useful adjunct in clinching an accurate diagnosis, as was seen in this case*

THANK YOU