CASE REPORT

Multiple Retroperitoneal Schwannomas with Cystic Degeneration Masquerading as Disseminated Hydatid Cysts: Case Report

Anshu Gupta Devra, Monal Trisal , Jyoti Mishra

Abstract

Schwannomas are benign nerve sheath tumors arising from differentiated Schwann cells. Common location is head& neck region, they are generally solitary. Retroperitoneal schwannomas are relatively rare accounting for 0.3 - 3.2% of benign schwannomas. Multiple shwannomas in the retroperitoneal region are extremely rare with only 4 cases reported till date. Pre-operative diagnosis of retroperitoneal schwannomas is difficult because they remain asymptomatic and also because they have a tendency to undergo cystic degeneration causing confusion in imaging studies. We present a case of 45 year old lady operated with provisional diagnosis of disseminated retroperitoneal hydatid cysts , which after histopathological examination was confirmed to be a case of multiple retroperitoneal schwannomas.

Keywords

Retroperitoneal Schwannoma, Cystic, Multiple

Introduction

Schwannomas are benign nerve sheath tumours arising from differentiated schwann cells. They are generally solitary, encapsulated tumours usually found in the head& neck region and on the flexor surfaces of the extremities. They show slight female predominance occurring between 2-5 decade. [1] Retroperitoneal schwannomas are relatively rare accounting for 0.3 - 3.2% of benign schwannomas. [2] More than one i.e., multiple occurrences in the retroperitoneal region are extremely rare [3], till date only 4 cases have been reported in literature, ours being the 5th one. Diagnosis of retroperitoneal schwannomas is difficult because they remain asymptomatic or present with vague clinical manifestations. Sometimes they exert pressure on adjacent structures or nerves bringing these tumours to clinical attention.[4] Preoperative imaging diagnosis is also tricky because of its deep location and lack of specific features on both ultrasound and computed tomography (CT). We are hereby reporting a rare case of 45 year old lady operated on the basis of provisional diagnosis of disseminated hydatid cyst on account of its imaging findings of multiple cystic lesions which subsequently after histopathological examination of the specimen submitted turned out to be multiple shwannomas with cystic degeneration.

Case Report

A 45 years old postmenopausal lady presented in medicine OPD with a complain of pain in abdomen and hematuria.

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Fig 1.A Gross Examination - Multiple Nodular Mass, with Cystic Degeneration (arrow)

She had past history of hysterectomy for fibroid. Pre-operative evaluation done revealed her liver function tests within reference range. Kidney function tests revealed mildly raised urea and creatinine. Her complete blood counts and differential count were within normal limits. Whole abdomen USG imaging showed multiple well-defined lesion, largest one measuring 4.4x3.9cm in paraaortic area, another in prevesicle area measuring 1.8x1.4cm and smallest one in left iliac fossa region. A presumptive diagnosis of multiple hydatid cysts was entertained.

Computed Tomography-abdomen revealed evidence of multiple retroperitoneal well defined well to lobulated heterogeneously enhancing round to oval hypodense lesion with multiple internal septations. An impression of multiple round to oval enhancing lesions in the retroperitoneum, pelvis and left sided anterior abdominal wall suggestive of hydatid cysts was given.

Patient underwent laparotomy and multiple swellings from parietal area, anterosuperior iliac spine, suprapubic, prevesical area and enlarged lymph nodes along with gall bladder was excised and was submitted for histopathological examination.On gross examination, excised specimen consisted of multiple globular to irregular greyish white soft tissue pieces varying in diameter from 1 to 4 cm. Cut surface of all the masses was homogenous with some showing cystic changes and foci of necrosis (***Fig 1***)

Histopathological examination from all of them revealed well encapsulated spindle cell tumor with cells arranged

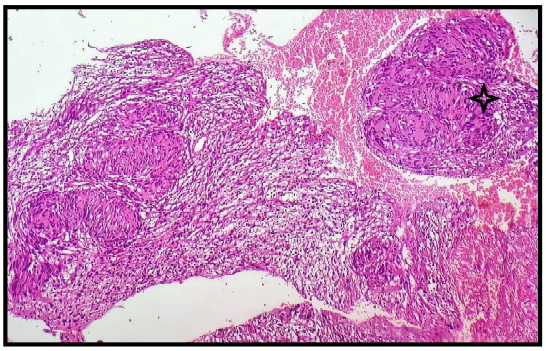

Fig 2 . Shwannoma with hypocellular and hypercellular areas-verocay bodies (star) [H&E X200]

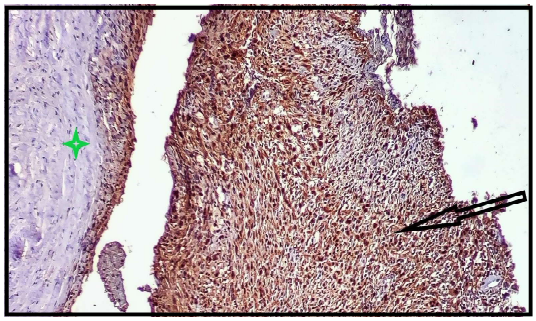

Fig 3. Tumor cells showing nuclear S-100 positivity (arrow) with unstained capsule at periphery (star) [IHC X 400)

in fascicles. There were both hypocellular and hypercellular areas with occasional verocay body formation, hyalinized thick walled blood vessels and areas of secondary xanthogranulomatous changes. Mitosis were inconspicuous. No atypia/ malignant changes were seen in the sections examined. Degenerative changes in the form of cystic degeneration, coagulative necrosis could be seen. Immunohistochemical staining for S-100 was diffusely and intensly positive confirming nerve sheath origin (***Fig 2&3***)

Based on the above findings diagnosis of peripheral nerve sheath tumor- Schwannoma was rendered. Also in view of multiple sites of involvement, patient was advised to be screened for Neurofibromatosis type 2.

Discussion

Schwannomas (neurilemmoma) are benign nerve sheath tumours that usually show predilection for head, neck and the flexor surfaces of the extremities. They are rarely seen in the retroperitoneal cavity accounting for 0.3-3.2% of schwannomas. [2]

Retroperitoneal tumours constitute a difficult management

problem due to their anatomic location, late presentation and proximity to adjacent vital structures thus making resection difficult or even impossible. They often become large in size due to the presence of loose areolar tissue in the retroperitoneum, and vague clinical symptoms leading to a delay in diagnosis. Common symptoms being vague, poorly localised pain and discomfort. Atypical presentations are very rare and include flank pain, haematuria, headache, secondary hypertension and recurrent renal colic pain.[5] Retroperitoneal schwannomas show cystic degeneration in up to 60% of cases while calcification is seen in 23% of cases only making diagnosis challenging especially radiologically. [6] Although radiological features of schwannoma such as target and fascicular signs are characteristic, these are infrequently seen in retroperitoneal schwannomas.

Schwannomas are encapsulated tumours. Microscopically, they demonstrate Antoni A areas (densely cellular, arranged in short bundles or interlacing fascicles) and Antoni B areas (fewer cells, organized, with great myxoid component). Long standing (ancient) shwannomas undergo degeneration due to central tumour necrosis as it out grows its blood supply. Malignant change in schwannomas is extremely rare, but when present, acts as high grade sarcomas producing local recurrence and distant metastasis. Features pointing towards malignancy are high mitotic index, pleomorphism and lymph vascular infiltration. Multiple retroperitoneal schwannomas are exceedingly rare. On searching available literature we could find till now only 4 reported cases of multiple retroperitoneal schwannomas [7]. Li *et al*. [8]

reported 82 cases of retroperitoneal schwannomas, 2 of which were multiple schwannomas (2.4%). The other 2 reports were case reports [9] making ours 5th reported case in literature. The differential diagnosis of such cystic lesion in retroperitoneal region includes pancreatic cystic tumours, hepatic tumours, psoas abscess, liposarcomas, and hemangiopericytomas. But characteristic histopathological features (Antony A & B areas) can clinch the diagnosis in majority of cases. Our patient had multiple benign neurilemmoma/schwannoma with cystic degeneration making it even more rarer as most of the cases reported in literature are solitary in nature. Multiple lesions associated with cystic degeneration resulted in its getting confused with disseminated hydatid cyst on imaging and clinical presentation. One such case misdiagnosed as hydatid has also been reported by Sahoo *et al* [5] but the lesion was solitary in nature. Treatment of such case is wide excision followed by careful monitoring as few cases have shown metastasis even after resection of histologically benign shwannoma.[6]

Conclusion

Retroperitoneal shwannomas although rare, can pose diagnostic dilemma on account of their propensity to undergo cystic change and this problem can be further compounded if they are multiple as happened in our case where on account of it being multiple and cystic, it was confused with disseminated hydatid cysts and therefore should be kept in differential diagnosis of multiple retroperitoneal cystic lesions.

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