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A Case of Primary Retroperitoneal Synovial Sarcoma

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A retroperitoneal Synovial Sarcoma is defined as a tumor arising in the retroperitoneal space, with an origin of mesodermal structures. The majority of synovial sarcomas (85 to 95%) occur in the extremities, located near the large joints. They are uncommon, and usually appear as a non-specific soft tissue mass, with no specific imaging feature. However, in some cases, radiological findings and anatomic location of the tumor may help the diagnosis. On CT examination, these tumors may be shown as hypo-dense masses and have an irregular enhancement in the periphery, with a poor enhancement in the central area, reflecting the necrotic, cystic and hemorrhagic changes. Histologically, there are two types of synovial sarcoma: biphasic and monophasic. Both types have about a 40% mortality rate. The known prognostic factors are; frequent mitotic figures, extensive tumor necrosis, and poor differentiation. Surgical ablation remains the mainstay of their management. Regardless of adequate surgical and adjunctive therapies, the recurrence rate ranges from 28 to 36%. Here, we present a case of a retroperitoneal synovial sarcoma, with a review of the literature. (J Korean Surg Soc 2003;64:256-260)

Key Words: Retroperitoneal neoplasm, Synovial sarcoma

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활막 육종은 원시 중간엽 세포기원의 악성 종양으로 모양이 활막 세포와 비슷하여 활막 육종이라 불리는데, 관절, 활액낭, 건초 부위에서 주로 생기며 약 5~15%는 두경부, 종격동, 복벽, 후복막에서 발생한다. 활막 육종은 젊은 나이(15세 이하)에 발생하거나 종양의 크기가 5 cm 이하일 경우 비교적 예후가 좋지만, 후복막에서 발생하는 경우는 매우 드물며 예후 또한 좋지 않다. 활막 육종은 방사선검사에서 다른 연부조직 종양과 구별되는 특징적인 소견이 없는 경우가 많으며 복부 전산화 단층 촬영에서는 일부에서 종괴내 괴사성 또는 출혈성 변화가 있으며 약 30%에서 종괴내 석회화나 골화가 발견되는 경우가 있다. 조직학적으로 단상성 활막 육종과 이상성 활막 육종으로 구별할 수 있으며 단상성 활막 육종이 이상성 활막 육종보다 예후가 좋다고 한다.(15) 활막 육종의 예후는 환자의 나이, 종양의 크기, 종양의 분화도 및 침투 정도에 의해 결정되며, 일부 유전자의 활성화도나 표현에 의해 영향을 받기도 한다고 한다.(12) 활막 육종의 치료 원칙은 수술적 완전 절제이며 추가적인 항암 치료나 방사선 치료는 육종의 재발을 줄이기 위해 수술 후 사용되나 생존율 향상에는 기여하지 못한다.

저자들은 63세 남자에서 발생한 원발성 후복막 활막 육종 1예를 경험하였기에 문헌 고찰과 함께 보고하는 바이다.

: 63세, 남자

: 내원 10여일 전부터 우하복부에 통증을 동반한 종괴를 주소로 타 병원에서 일차 검진 후 우측 대장암의 의심되어 본원 외래를 방문하였다.

: 특이사항 없음, 최근 복부에 외상받은 적도 없었음.

가 : 특이사항 없음.

: 내원시 혈압은 110/60 mmHg, 맥박수 60회/분, 호흡수 20회/분, 체온은 36.2°C이었고, 두경부 및 사지 특이 소견은 없었으며, 폐음 및 심음도 정상이었다. 우측 하복부에 약 8×6 cm 크기의 압통을 동반한 단단한 종괴가

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13.1 g/dl, 37.6%, 10900/mm³, 234 K/mm³

CEA α P

10 cm

가 (Fig. 1).

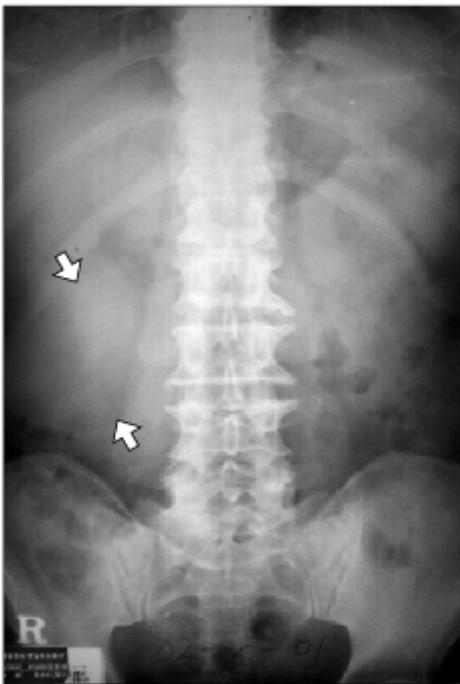


Fig. 1. Plain film of the abdomen showed a large ovoid homogenous opacity mass (arrow) in the Rt. upper abdomen.
가 가

(Fig. 2),

10×8 cm

가 , 가

가

(Fig. 3A),

(Fig. 3B).

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(Fig. 4).

: 10×8 cm



Fig. 2. Ultrasonogram showed a large mixed echoic mass in Rt. intraabdominal cavity.

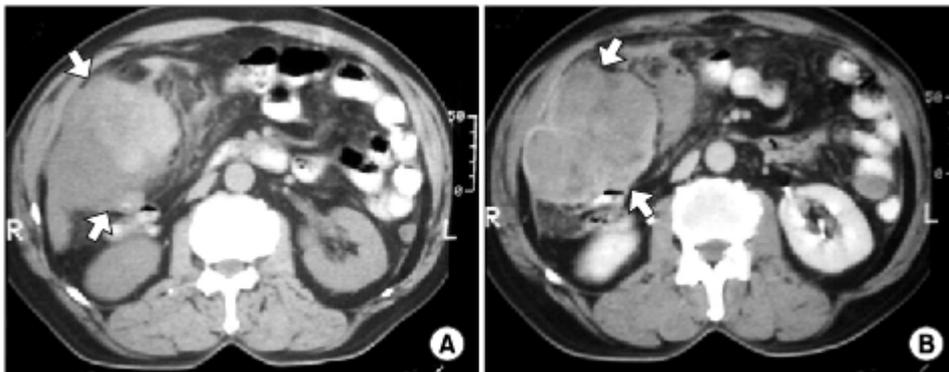


Fig. 3. (A) Pre-contrast enhanced computed tomogram (CT) revealed a 10×8 cm lobulate, marginated, heterogenous, attenuating mass (arrow) with internal high attenuated region, reflecting the hemorrhagic foci. (B) Post-contrast enhanced CT scan revealed moderate heterogeneous enhancements (arrow) with moderate peritumoral infiltrations and ascites.

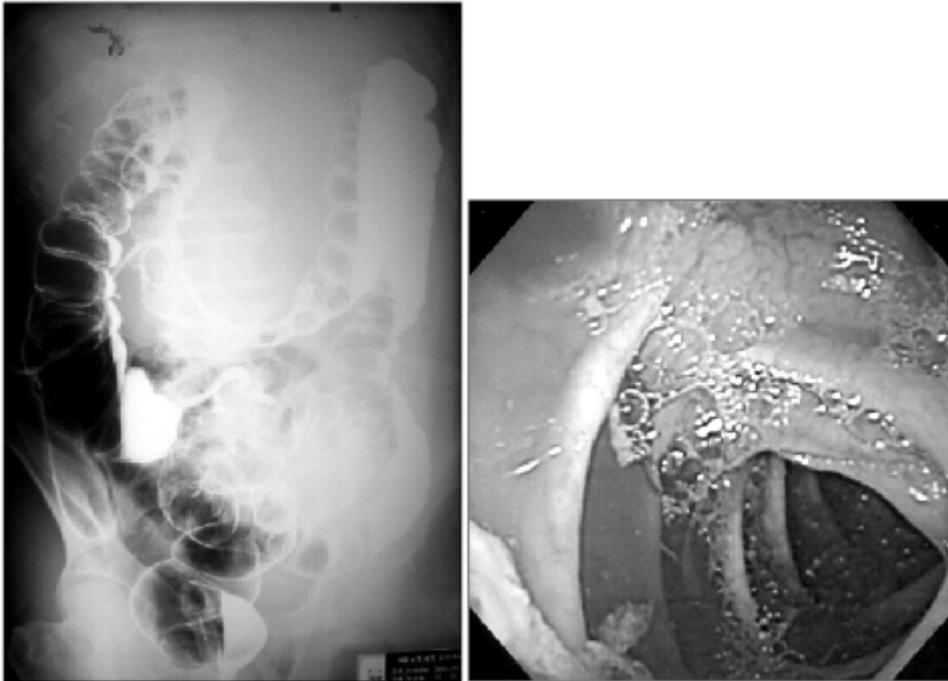


Fig. 4. Barium enema and colonoscopic examination showed normal mucosal pattern.



Fig. 5. The cut-surface has a fish fresh gray-tan appearance with hemorrhage.

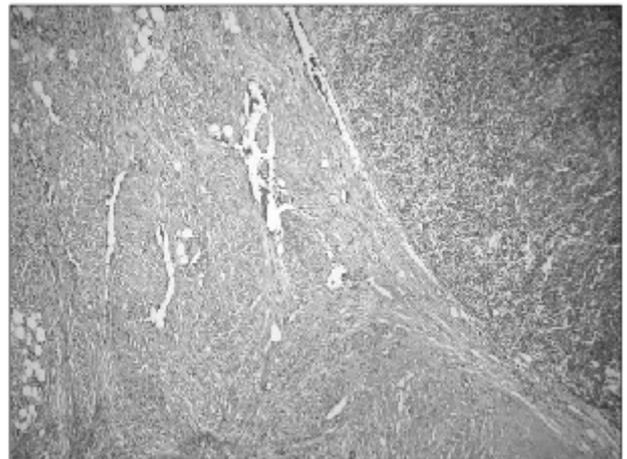


Fig. 6. Biphasic synovial sarcoma with glandular epithelial structures surrounded by spindle cell element (H&E stain× 100).

1×1 cm 가
 : 가
 가 13×11×8 cm
 가

(Fig. 5).

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 15
 (Fig. 6).
 cytokeratin epithelial membrane antigen (EMA)
 vimentin

(Fig. 7).

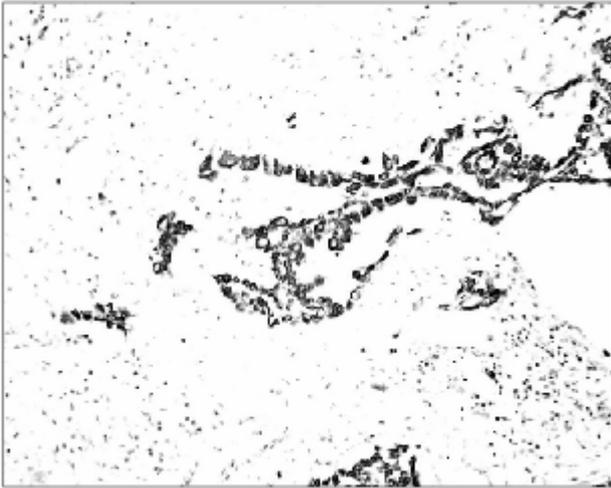


Fig. 7. High molecular weight cytokeratin immunoreactivity highlight the epithelial elements in the biphasic synovial sarcoma (x200).

가 .(1,4)
가 ,
(epithelioid cell) (spindle cell)가
(Biphasic) , 가 가
(Monophasic)
(3-5,8) E-cadherin
가
(cytokeratin), (epithelial mem-
brane antigen) bcl-2 가
S1C CD99(MIC2)
(t(x;18)(p1.2;q11.2))
X 가
SSX SYT
(9,10)
(fibrosarcoma) ,
(hemangiopericytoma),
(malignant peripheral nerve sheath tumor; MPNST, malignant schwannoma)
(cytokeratin) ,
가 ,
S-100
(11)
(≤5 cm vs >5 cm), ,
가 p27
APC
APC
(Missense mutation) β-
catenin β-catenin
(oncoprotein)
(12-14)
HPF 10 ,
(15)
30~70% 2
가 18%
가
74~81%, 12~23%,

:
11
5~10% ,
15~40 ,
X
(1,2)
80~95% , ,
(3-5)
가
가 가
가
가
(6,7)
가 가

10~20% 가 가 .(1,15)
 25 , 5 cm 가
 , 3.6
 .(16)
 가 가 ,
 .(7)
 (doxo-
 rubicin) (ifosfamide) 24%
 ,
 .(17)

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