Mahidol University
Faculty of Medicine
Siriraj Hospital

SIENDO
Faculty of Medicine Siriraj Hospital

Interhospital conference
Case 2

24th March 2023

F1 Benjawan Soontornrungsun

Asst. Prof. Taweesak Wannachalee

Teratoma

- Teratomas are originating from embryonal germ layers, the majority of them are mature, which are benign
- Incidence 0.9/100,000
- Most common in the gonads, only 15% out of gonads
- Primary adrenal teratomas are extremely rare accounting for 0.7% of all primary adrenal tumors

Wang et al. Front. Oncol. 12:830003

Teratoma

Criteria for benign teratomas

- Absence of malignant or immature elements in histopathology
- Absence of metastasis
- Normal levels of AFP and hCG
- Absence of recurrences during follow-up

Li et al. World J Surg Onc 13, 318 (2015)

Criteria for malignant teratoma

- Immature teratoma
- Teratoma with other malignant germ cell tumor components (such as yolk sac tumor, embryonal carcinoma, choriocarcinoma, and seminoma)
- Teratoma with malignant transformation (TMT)

Morinaga et al. Am J Clin Pathol 101: 531-534

Malignant teratoma common in 15 - 35 year old males

Einhorn et al., 1985

Teratoma with Malignant Transformation (TMT)

- Extremely rare, most are found in gonadal tumors
- Probably by partial differentiation of totipotential germ cells to somatic tissue with concomitant malignant transformation, or by malignant transformation of pre-existing teratomatous elements

Ulbright et al. Cancer 54: 1824-1833

Adrenal Teratoma

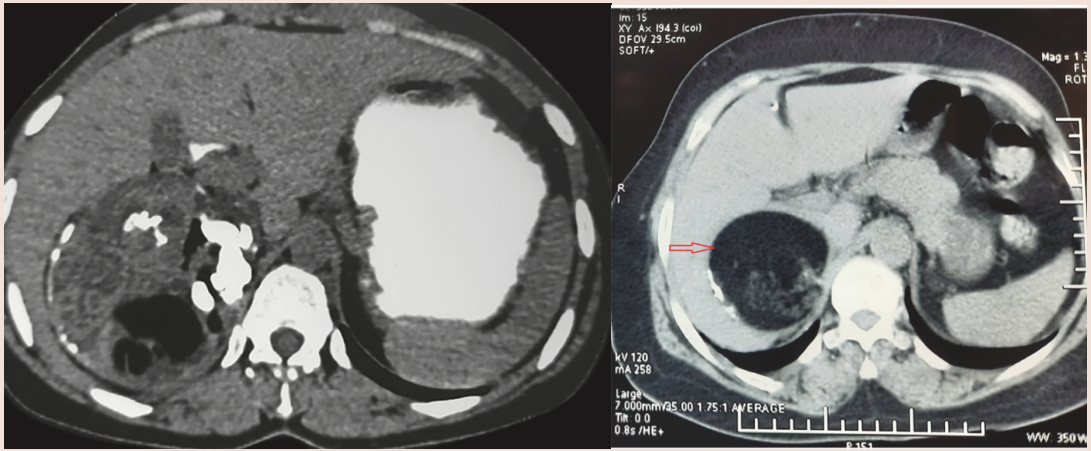
- Usually asymptomatic, laboratory tests are generally normal
- Usually large, as the tumors grow, low back pain and paroxysmal abdominal pain may occur
- Ultrasound: heterogeneously echogenic mass including hyperechoic calcifications and ossifications, as well as hypoechoic fatty and cystic areas
- CT scan: Large heterogeneous density with hypointense cystic and fatty areas and hyperintense calcifications is characteristic

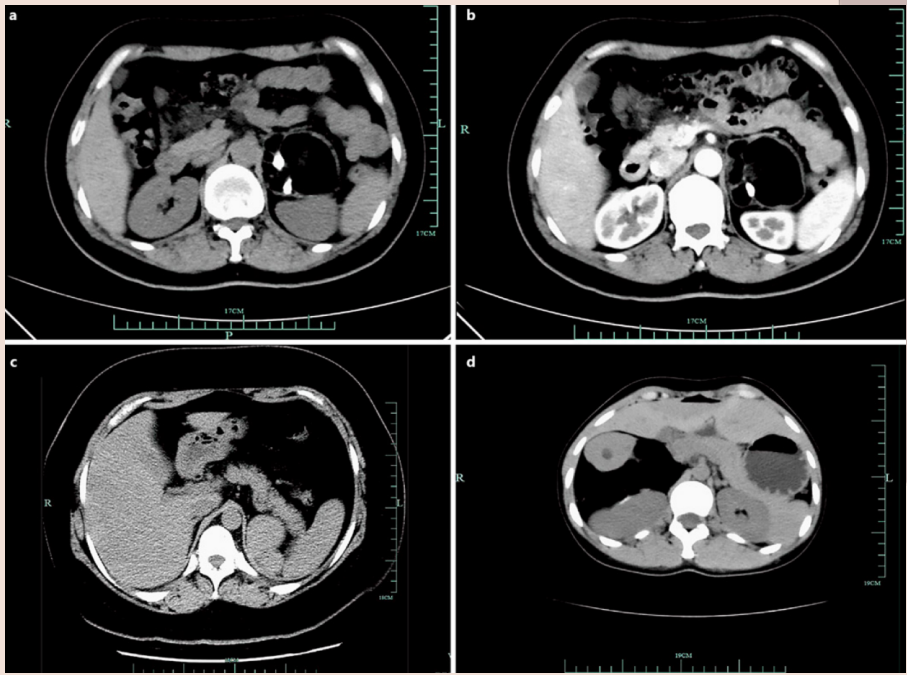
Elevated AFP, β-HCG, and CEA were found in some immature teratomas

d'Amuri et al. Gland Surg (2019) 8(Suppl 3):S223–S32

Lam et al. Surgery 1999;126:90–4

Adrenal Teratoma



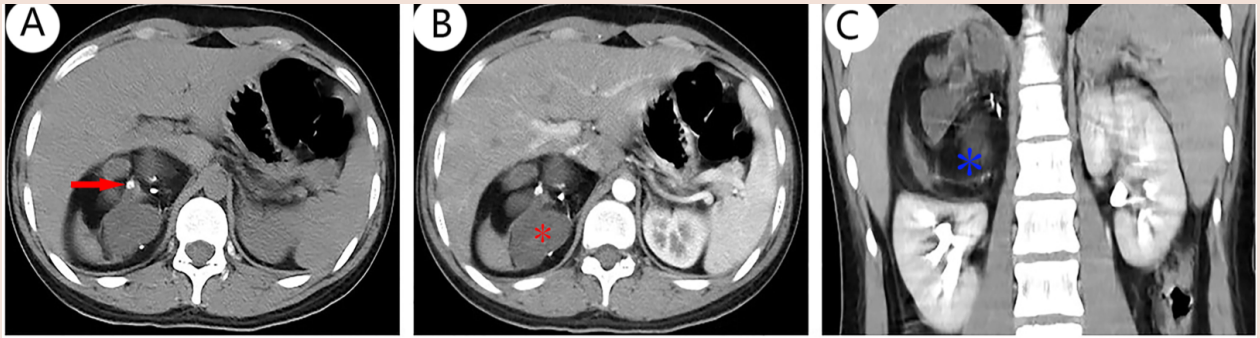


Primary adrenal teratoma mean diameter 8.25 cm

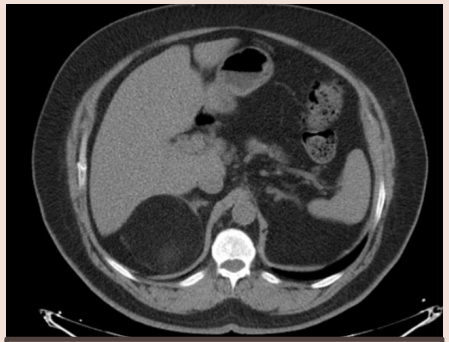
Ramakant et al. J Postgrad Med (2018) 64(2):112–4

GUO et al. Eur. J. Radiol. 62 (2007) 359–370

Adrenal Teratoma

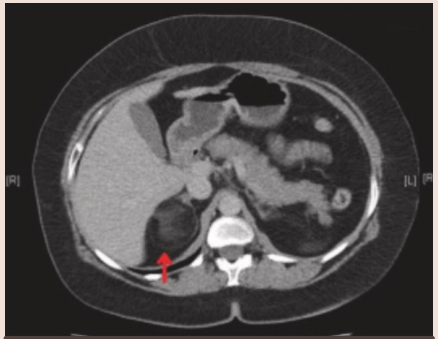


Well-circumscribed calcification and contained fat components were important signals for the diagnosis of adrenal teratoma



adrenal myelolipoma

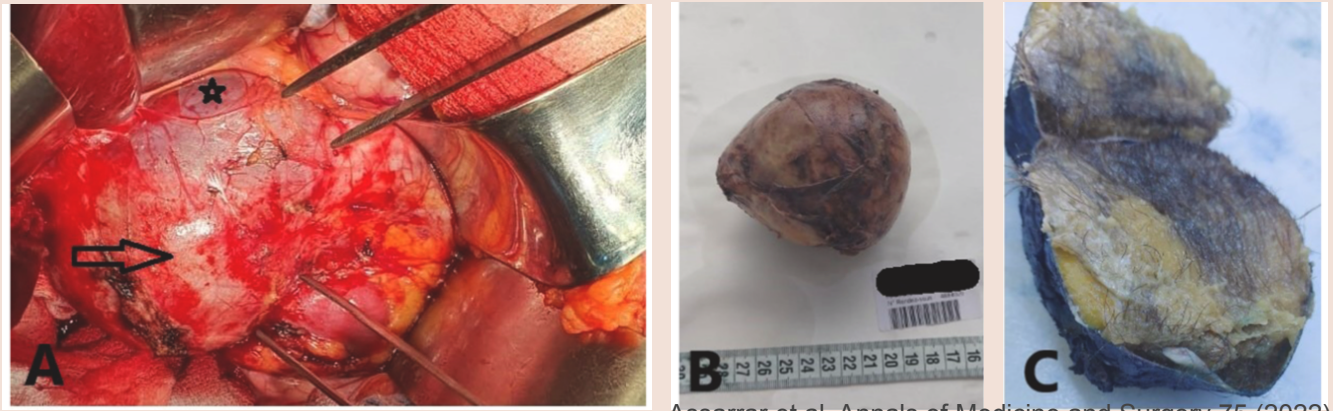
adrenal angiomyolipoma



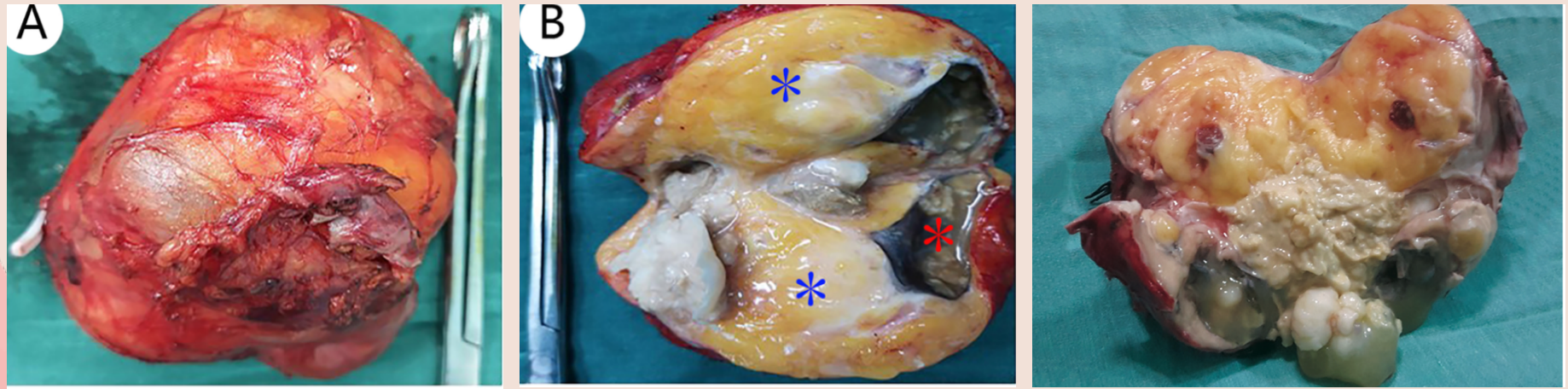
Adrenal lipoma

Differentiate adrenal teratoma from common adrenal lipoma diseases, such as adrenal myelolipoma, adrenal angiomyolipoma, lipoma, gangliocytoma

Adrenal Teratoma



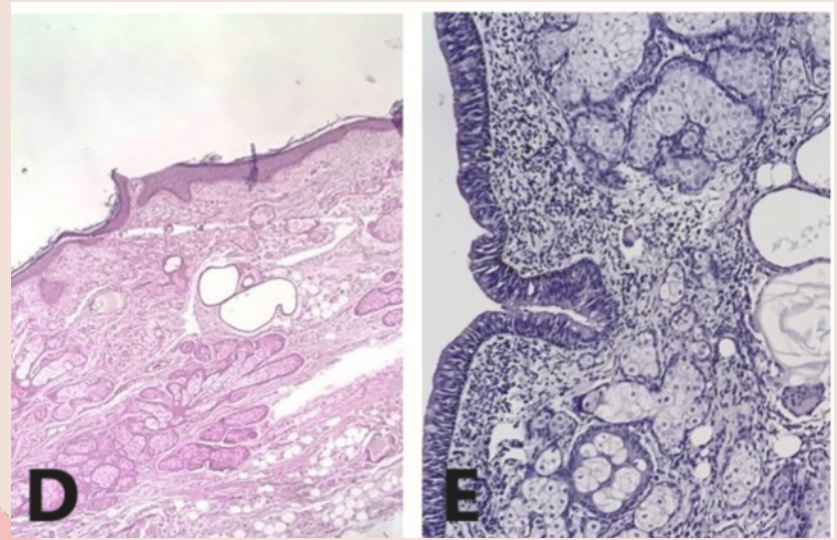
Assarrar et al. Annals of Medicine and Surgery 75 (2022)



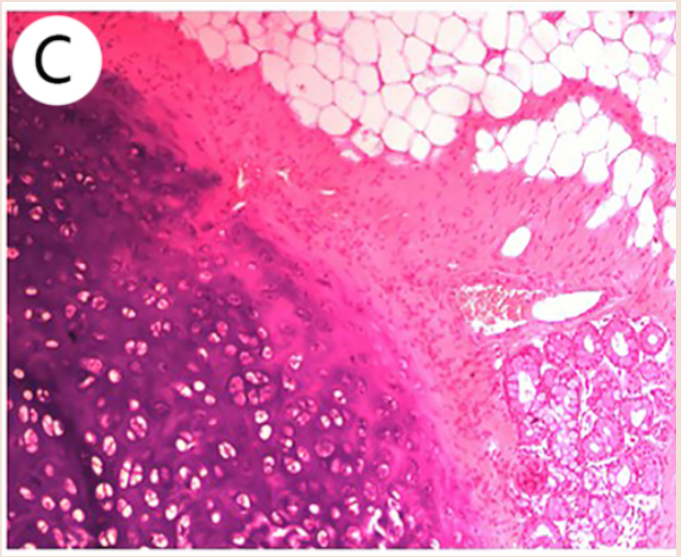
Wang et al. Front. Oncol. 12:830003

Adrenal Teratoma

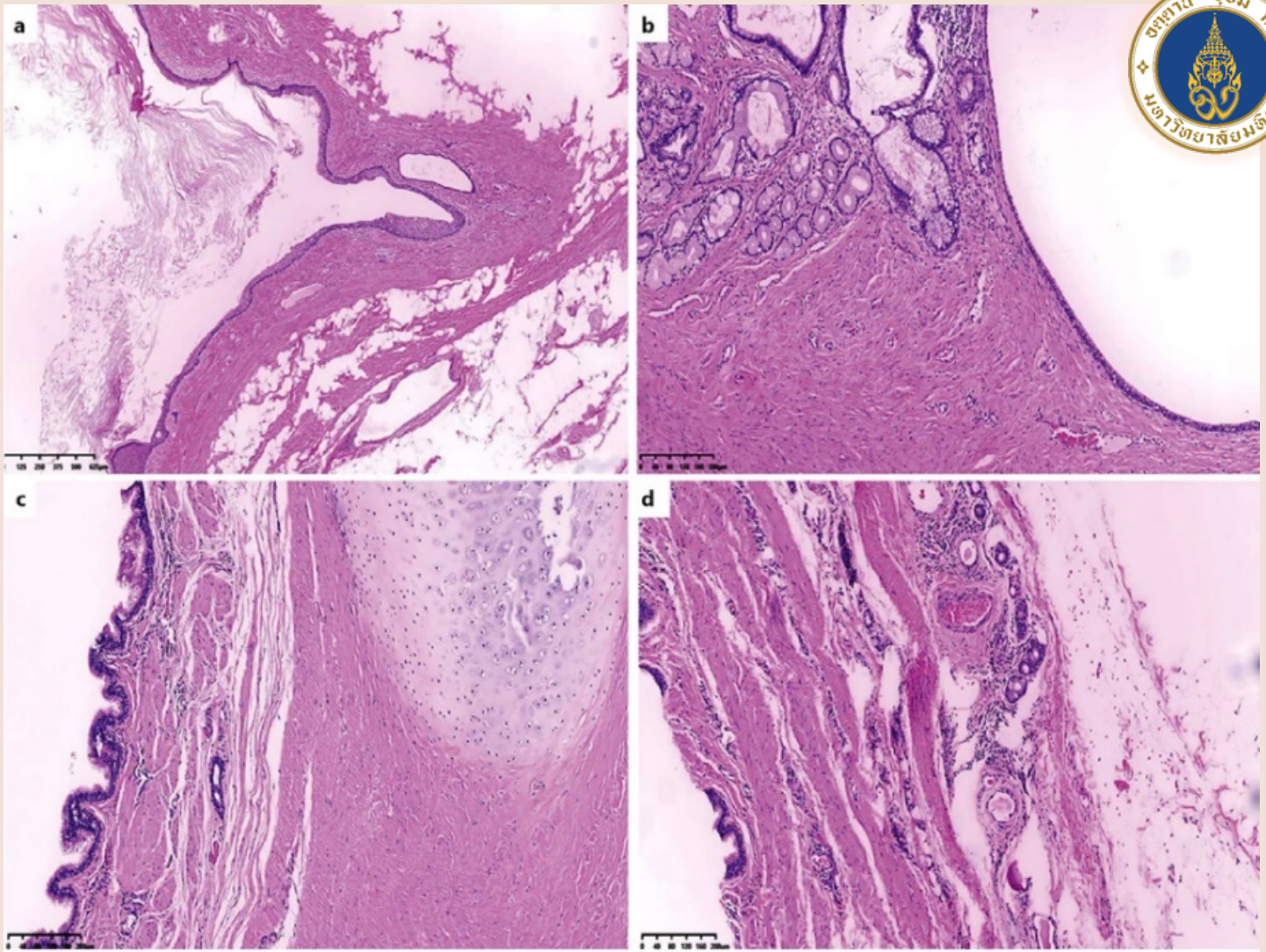
- Histopathological examination is the key for a conclusive diagnosis
- Mature teratomas appear as well-encapsulated tumors, mainly cystic and composed of two or three germ layers, usually comprising fully mature elements i.e., sebaceous material, hair, bone, and fat components
- Immature teratomas include at least 10% of undifferentiated tissue



Assarrar et al. Annals of Medicine and Surgery 75 (2022)



Wang et al. Front. Oncol. 12:830003



Urol Int 2022;106:1195–1199

Treatment and Prognosis

- Surgery should be performed as soon as possible

  For relief symptoms related to the mass and teratoma have the potential for malignancy

- Prognosis after resection of mature teratomas is good

  Overall 5-year survival rate nearly 100%

- Teratoma with malignant transformation

  -Malignant teratoma is prone to recurrence and requires adjuvant radiotherapy and chemotherapy with a lifelong follow-up

  -Good prognosis if tumor is completely excised and not extended beyond capsule

  - At present, no sensitive indicators for monitoring relapse of adrenal teratoma
  - The level of AFP was correlated with the recurrence of teratoma, and it can be used as a predictive index for curative effect

  Nam et al. J Pediatr Surg (2018) 53(12):2470–4

frontiers | Frontiers in Oncology

CASE REPORT
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Rare Primary Adrenal Tumor: A Case Report of Teratomas and Literatures Review

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- 49 patients (14 men, 35 women)
- Mean age of 30 years
- Adrenal-related hormones were normal in all patients
- Clinically asymptomatic 55%
- The majority were unilateral, right-sided, and mature teratomas
- One was a malignant transformation of a teratoma into an adenocarcinoma
- 39 of the 49 patients followed up for 3 months to 10 years, and all had a good prognosis with no tumor recurrence

TABLE 2 | Review of characteristics of adrenal teratoma.

Cas No.	First author, year	Age (years) /Sex	Symptoms	Location	Size(cm)	Pathology	surgery	follow-up (months)
1	Li et al., 2011 (10)	4/F	Asymptomatic	Left	3.0×2.0×2.0	MT	OA	24
2	Li et al., 2011 (10)	38/F	Right waist pain after fatigue	Right	11.0×8.0×6.0	MT	OA	24
3	Zhao et al., 2014 (11)	21/F	Backache on the right side	Right	6.0	MT	OA	80
4	Zhao et al., 2014 (11)	35/F	Asymptomatic	Right	8.0	MT	OA	57
5	He et al., 2020 (12)	17/F	Asymptomatic	Right	7.0×2.5×2.0	MT	LA	12
6	Ban et al., 2019 (13)	60/M	left flank pain	Left	12.0×11.0×11.0	MT	NA	NA
7	Lam et al., 1999 (14)	18/F	Back pain	Left	11.0×8.0×7.0	MT	NA	84
8	Lam et al., 1999 (14)	17/M	Back pain	Right	7.5×6.0×3.0	MT	NA	6
9	Lam et al., 1999 (14)	37/F	Back pain	Left	10.0	MT	NA	96
10	Narla et al., 2016 (15)	2.5/F	right upper quadrant abdominal pain	Right	6.0×5.0×3.0	MT with carcinoid tumor	NA	8
11	Haddad et al., 2020 (16)	1/M	Asymptomatic	Left	12.0×10.9×8.5	MT	NA	6
12	Li et al., 2015 (17)	21/F	Asymptomatic	Right	8.5	MT	LA	NA
13	Li et al., 2015 (17)	16/M	Asymptomatic	Right	9.0	MT	LA	NA
14	Li et al., 2015 (17)	43/F	Asymptomatic	Left	4.9	MT	LA	NA
15	Li et al., 2015 (17)	49/F	Asymptomatic	Left	5.3	MT	LA	NA
16	Li et al., 2015 (17)	51/F	Asymptomatic	Right	2.4	MT	LA	NA
17	Zhou et al. 2018 (18)	69/F	Asymptomatic	Left	10.0×6.0×4.0	MT	NA	12
18	Zhou et al., 2018 (18)	29/F	Asymptomatic	Left	2.5×2.1×0.5	MT	LA	12
19	Ramakant et al., 2017 (19)	25F	right hypochondriac pain	Right	19.0×15.0	MT	OA	NA
20	Ersoz et al., 2011 (20)	8/M	right side pain during exercise	Right	10.0×8.0×6.0	neurocytoma arising in MT	OA	6
21	Niu et al., 2017 (21)	36/F	Asymptomatic	Right	9.0×6.0×7.5	malignant transformation of MT	LA	NA
22	McMILLAN et al., 1987 (22)	17/M	left loin pain	Left	8.0	undifferentiated MT	NA	NA
23	Wang et al., 2019 (23)	22/F	Asymptomatic	both	left:10×10.8×12.9; right:10.3×12.3×12.1	MT	LA	10
24	Castillo et al., 2006 (24)	8/M	lumbar pain after a fall	Right	8.0	MT	LA	36
25	Castillo et al., 2006 (24)	61/F	Asymptomatic	Left	8.0	MT	LA	12
26	Li et al., 2015 (25)	49/M	Asymptomatic	Right	6.0×7.0×11.0	MT	LA	8
27	Ciftci et al., 2013 (26)	0.25/M	Asymptomatic	Left	8.3×8.0	MT	NA	NA
28	Pandit et al., 2018 (6)	16/M	Asymptomatic	Left	12.0×10.0	MT	OA	12
29	Zhong et al., 2019 (27)	59/F	Flank pain	Left	10.6	MT	OA	124
30	Zhong et al., 2019 (27)	54/F	Asymptomatic	Right	10.2	MT	OA	124
31	Zhong et al., 2019 (27)	22/F	Abdominal pain	Right	10.0	MT	OA	109
32	Zhong et al., 2019 (27)	48/F	Asymptomatic	Right	9.6	MT	LA	101
33	Zhong et al., 2019 (27)	26/M	Flank pain	Right	4.0	MT	LA	90
34	Zhong et al., 2019 (27)	18/F	Abdominal pain	Right	8.8	MT	LA	88
35	Zhong et al., 2019 (27)	55/F	Asymptomatic	Left	14.0	MT	OA	69
36	Zhong et al., 2019 (27)	28/F	Asymptomatic	Right	4.5	MT	LA	60
37	Zhong et al., 2019 (27)	29/F	Asymptomatic	Right	7.8	MT	LA	56
38	Zhong et al., 2019 (27)	29/F	Asymptomatic	Left	7.0	MT	LA	56
39	Zhong et al., 2019 (27)	72/F	Asymptomatic	Right	6.0	MT	OA	55
40	Zhong et al., 2019 (27)	28/F	Asymptomatic	Left	9.5	MT	OA	76
41	Zhong et al., 2019 (27)	41/F	Asymptomatic	Left	18.0	MT	OA	43
42	Zhong et al., 2019 (27)	45/M	Asymptomatic	Left	6.8	MT	LA	28
43	Bhatia et al., 2016 (28)	24/F	left hypochondrium pain	Left	7.6×6.5	MT	NA	NA
44	Kuo et al., 2017 (29)	26/M	right upper quadrant and right flank pain	Right	8.5×5.5×4.8	MT	OA	NA
45	Kuo et al., 2017 (29)	29/F	Asymptomatic	Left	2.5×2.1×0.5	MT	LA	NA
46	Kuo et al., 2017 (29)	24/F	left lower quadrant abdominal pain	Left	11.5×9.0×3.0	MT	LA	NA
47	Garg et al., 2017 (30)	0.25/F	Asymptomatic	Right	10.0×10.0×8.0	MT	OA	NA
48	Tang et al., 2014 (31)	39/F	dizzy	Right	22.5×17.0×7.0	MT	NA	18
49	Nadeem et al., 2015 (32)	19/M	vague pain in right flank	Right	8.0×6.0×4.0	MT	OA	12

F, female; M, male; MT, mature teratoma; OA, open adrenalectomy; LA, laparoscopic adrenalectomy; NA, not available.

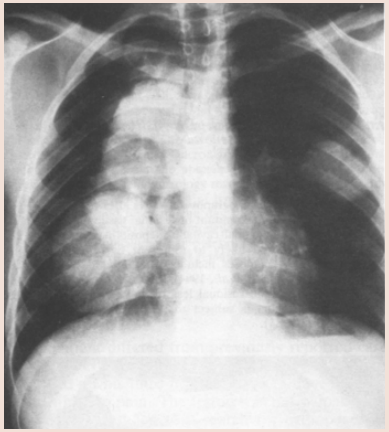
Clinical Radiology (1987) **38**, 327–328

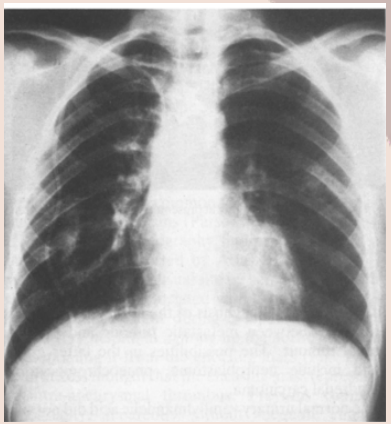
Case Report: Malignant Teratoma Presenting with an Adrenal Mass

A. McMILLAN and A. HORWICH

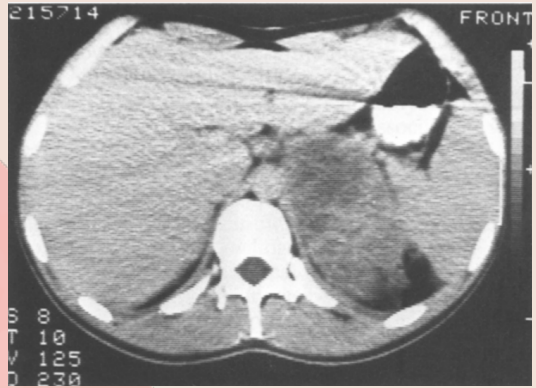
Department of Radiotherapy, Royal Marsden Hospital and Institute of Cancer Research, Downs Road, Sutton, Surrey

790 patients with malignant teratoma between 1970 - 1984,
5.7% were extragonadal in origin
75% of extragonadal malignant teratoma elevate serum AFP or β-hCG





17 year old man with 6 week history of shortness of breath, left loin pain and weight loss
Tender abdominal mass at left hypochondrium



- CT abdomen: 8 cm mass replacing the left adrenal gland, and para-aortic lymphadenopathy
- Chest radiograph: multiple large lung metastases
- Urinary VMA, serum AFP, and βhCG were normal
- Testicular ultrasound: normal
- Histological diagnosis by adrenalectomy: undifferentiated malignant teratoma
- Chemotherapy comprising cisplatin, bleomycin, vincristine, and etoposide

Clinical Case Report

Medicine

OPEN

Malignant transformation of a mature teratoma of the adrenal gland

A rare case report and literature review

Miao Niu, MD[a], Ailian Liu, MD[a,*], Ying Zhao, MD[a], Lu Feng, MD[b]

- 36-year-old Chinese female denied any symptoms
- Physical examination: normal

Cortisol ↑ , ACTH ↑ , Aldosterone ↑
24 h Urine VMA ↑
Angiotensin I and Angiotensin II were normal

Table 1

All hormonal test results of the patient.

Hormone name	Result	Reference range	Unit
Cortisol	773.58	118.6–618	mol/L
ACTH	70.98	7.1–63.3	pg/mL
Aldosterone	317.84 (Standing)	Prone: 49.3–175 Standing: 34.7–275	pg/mL
Angiotensin I	2.74 (Regular diet Standing)	Regular diet Regular diet pro: 0.05–0.79 Regular diet Standing: 0.93–6.56 Low sodium diet prone: 0.00–7.14 Low sodium diet Stand: 1.13–9.68	ng/mL/h
Angiotensin II	68.57 (Regular diet Standing)	Regular diet prone: 28.2–52.2 Regular diet Stand: 64.3–120.7 Low sodium prone: 40.6–91.0 Low sodium Standing: 64.3–120.7	ng/mL/h
24-h urine volume	2000	—	mL
VMA	22.38	0–13.6	mg/24 h

ACTH = adrenocorticotropic hormone, VMA = vanillyl mandelic acid.

Abnormal levels of hormones including cortisol, ACTH, aldosterone, and VMA may also aid in the diagnosis of adrenal TMT

Clinical Case Report

Medicine

OPEN

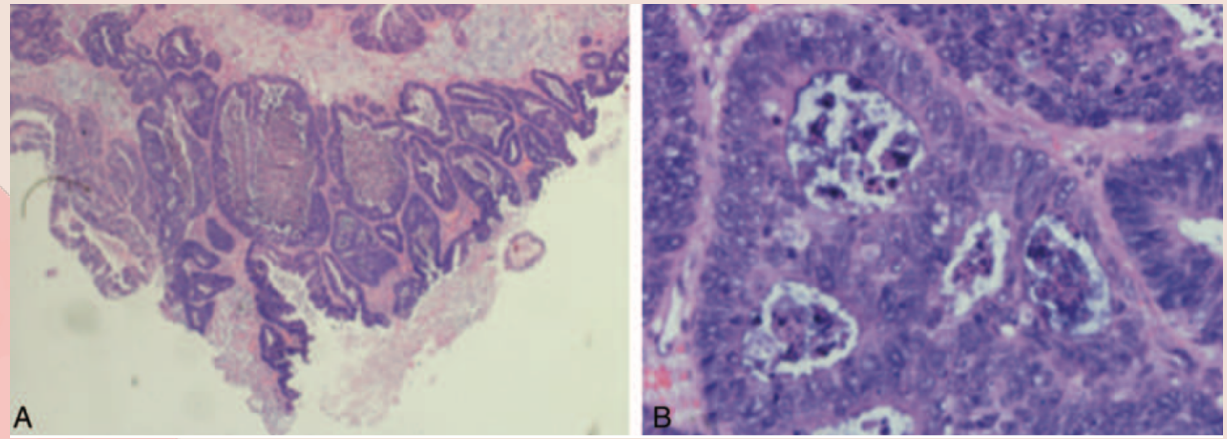
Malignant transformation of a mature teratoma of the adrenal gland

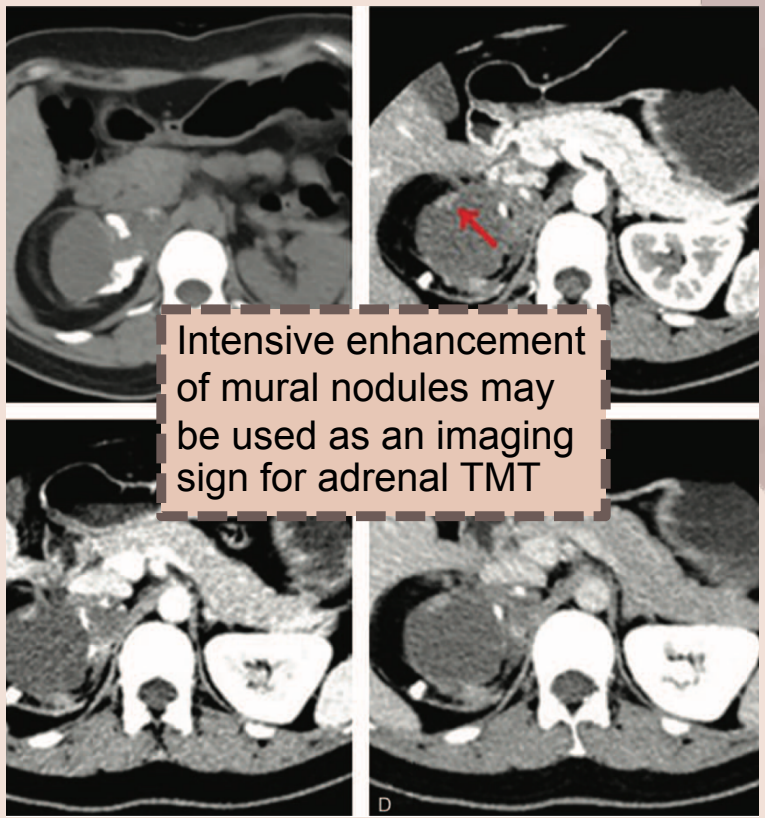
A rare case report and literature review

Miao Niu, MD[a], Ailian Liu, MD[a,*], Ying Zhao, MD[a], Lu Feng, MD[b]

Contrast-enhanced CT showed no significant enhancement in the low-density cystic and fatty areas, while the hyperdense peripheral rim and internal septations showed significant enhancement. Intense enhancement of mural nodules was noted

A well-defined mixed density mass 8 x7 x 6 cm in the right adrenal region. Hypodense areas dominated in the mass, with an even lower density fatty area along the right aspect of the mass. Patchy areas of high densities in the left aspect. The mass shows peripheral hyperdense rim and hyperdense internal septations





Intensive enhancement of mural nodules may be used as an imaging sign for adrenal TMT

Eur. Radiol. (2002) 12:1546–1549
DOI 10.1007/s003300101105

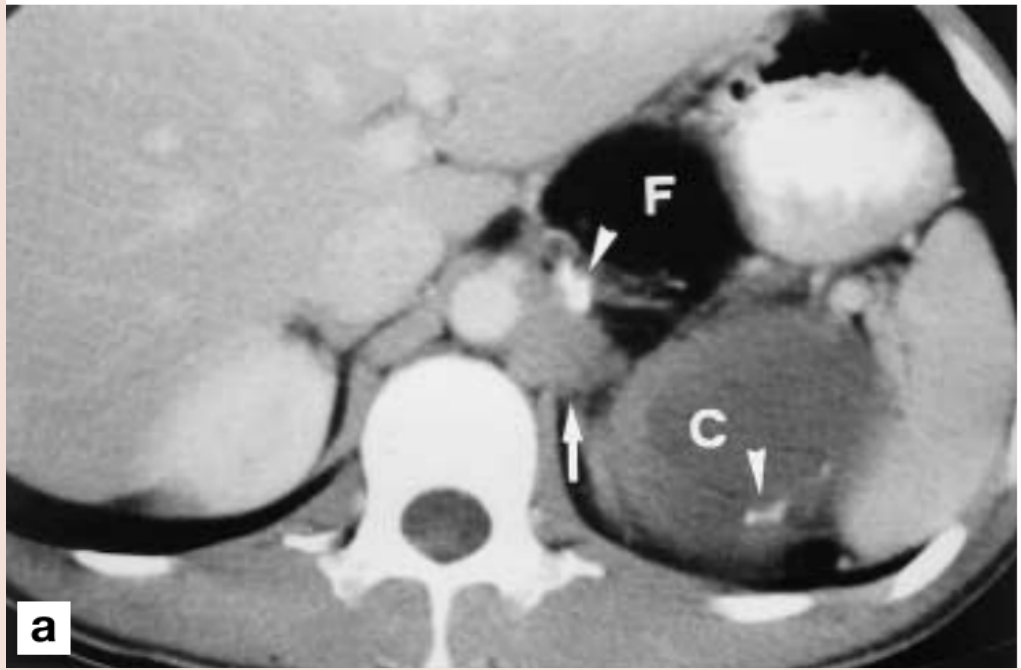
UROGENITAL

Li-Jen Wang
Sheng-Hsien Chu
Kwai-Fong Ng
Yon-Cheong Wong

Adenocarcinomas arising from primary retroperitoneal mature teratomas: CT and MR imaging

CT scan suggest TMT

- Irregular wall thickening of the cystic area, contiguous with the solid mass that involved the adjacent organs or lymph nodes



Conclusion

- Primary adrenal teratomas are extremely rare
- Usually asymptomatic and adrenal-related hormones were normal
- CT scan: large heterogeneous density with hypointense cystic and fatty areas and hyperintense calcifications is characteristic
- Differentiate adrenal teratoma from common adrenal lipoma diseases, such as adrenal myelolipoma, angiomyolipoma, and lipoma
- Surgery should be performed as soon as possible
- Prognosis after complete resection is good