EP.0322 - Success story of a girl presented with a massive yolk sac tumour

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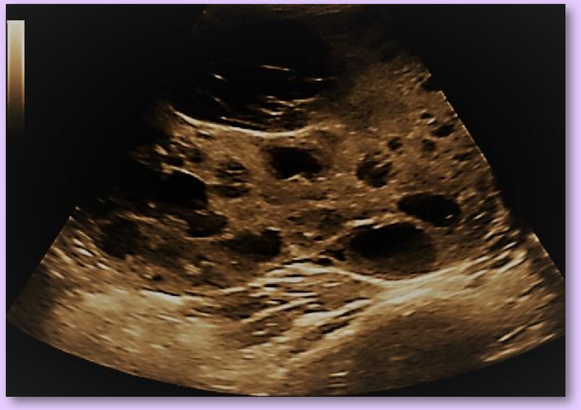
Objective

Among malignant ovarian germ cell tumors (MOGCT), yolk sac tumors (YST) are the second commonest and represent 20% of all MOGCTs. These aggressive tumors appear commonly in reproductive age group. With introduction of effective adjuvant chemotherapeutic regimens, fertility preservation is a realistic goal.

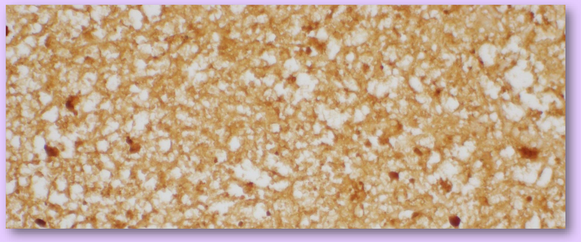
Case report

A 13-year-old otherwise healthy girl who attained menarche at 11years presented with a two-week history of fever, headache, dry cough, abdominal pain with recent onset loss of appetite and loss of weight. On examination she had an abdomino-pelvic mass compatible with a 34week gravid uterus. Ultrasonography revealed a large heterogeneous mass with cystic-solid regions, free fluids, right sided hydronephrosis and hydroureter. An ovarian origin was suggested by contrast enhanced computerized tomography (CECT). Distal spread was not noted radiographically. Pre surgical serum alpha-feto-protein (AFP) exceeded 5000IU/mL, carcinoma antigen125 was 391U/L and lactate dehydrogenase was 660U/L. An ovarian YST was predicted.

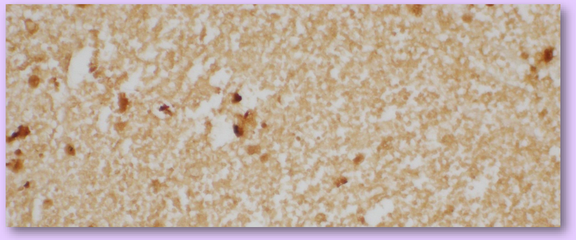
She underwent fertility-sparing debulking laparotomy with right salpingo-oophorectomy, since left adnexa was macroscopically normal. The surgical stage was FIGO 1C2. Histology confirmed a malignant YST without lympho-vascular invasion. Adjuvant chemotherapy was initiated following surgical recovery. She reached AFP level of 1.69IU/ml from immediate post-surgical level of 969IU/mL within 6months. She regained menstruation 4 months post-surgery and currently is on recommended follow up protocol.



Immunohistochemistry – strong AFP positivity



Immunohistochemistry – hCG negativity



Discussion

Most ovarian YSTs appear during second decade of life. Macroscopically they're firm, solid-cystic masses with predominantly solid components. It's invariably positive for AFP on immunohistochemical analysis. AFP is a reliable tumor-marker to determine the residual tumor, chemotherapy response and recurrence. Fertility sparing surgery is possible since 94% of tumors are unilateral. Preservation of ovarian tissue is considered even in bilateral disease. Adjuvant regimen of 3-4 cycles of BEP (bleomycin, etoposide, and cisplatin), which is recommended from stage I, was practiced in this case too. With surgery and adjuvant BEP, overall survival rate is 75%-90% for stage I & over 60% for advanced disease. The 10-year follow-up regimen includes clinical, radiological and tumor-marker assessments, with gradual increase between appointment intervals.

Conclusion

Age at diagnosis being less than 25 years, smaller primary tumor give better prognostic outcomes despite statistical insignificance. Early stage (I, II) disease, absent residual tumor, BEP regimen & rapid AFP depletion are established prognostic factors in YSTs. There's no added benefit of radical surgery over fertility prese`rvation in early-stage disease. The preoperative AFP value doesn't affect prognosis. Fertility success following completion of treatment exceeds 75%.

References

1. Cicin I, Saip P, Guney N, Eralp Y, Ayan I, Kebudi R, Topuz E. Yolk sac tumours of the ovary: evaluation of clinicopathological features and prognostic factors. Eur J Obstet Gynecol Reprod Biol. 2009 Oct;146(2):210-4. doi: 10.1016/j.ejogrb.2009.02.052. Epub 2009 May 5. PMID: 19419810.
2. Iscar T, Arean C, Chiva L, et al Ovarian yolk sac tumor International Journal of Gynecologic Cancer 2021;31:797-798.
3. Chen LH, Yip K-C, Wu H-J and Yong S-B (2019) Yolk Sac Tumor in an Eight-Year-Old Girl: A Case Report and Literature Review. Front. Pediatr. 7:169. doi: 10.3389/fped.2019.00169.
4. Sell A, Sogaard H, Norgaard-Pedersen B. Serum alpha-fetoprotein as a marker for the effect of post-operative radiation therapy and/or chemotherapy in eight cases of ovarian endodermal sinus tumour. Int J Cancer. 1976 Nov 15;18(5):574-80. doi: 10.1002/ijc.2910180505. PMID: 62720.