Choriocarcinoma Syndrome: A Lethal Sequelae of Testicular Tumor

Rajesh Batajoo, MBBS, MS; Atul Kasaju, MBBS, FCPS; Jagdish L Baidya, FRCS, FCPS
Department of Urology, B & B Hospital, Gwarko, Lalitpur, Nepal

Address for Correspondence:
Atul Kasaju, MBBS, FCPS
Department of Urology, B & B Hospital, Gwarko, Lalitpur, Nepal
Email: dratulkasaju1@gmail.com

Received, 16 May, 2018
Accepted, 25 May, 2018

A 25 years old gentleman presented with painless right scrotal swelling for 9 months. USG showed right testicular highly vascular solid mass with metastasis at the liver, right kidney and spleen. Tumor markers showed markedly raised β-hCG levels of 5,02,110 IU/L. Radical orchidectomy specimen was consistent with mixed germ cell tumor with 20% choriocarcinomatous component. CECT showed further metastasis at the brain and right lung. During the early post-operative period, there was bleeding from his kidney metastases causing huge perinephric haematoma leading to hemorrhagic shock. After patient was stabilized, chemotherapy was started which showed gradual improvement. However, patient again had intracerebral hemorrhage from metastatic site. Patient and his family refused further treatment, eventually the patient died after 2 months. Hemorrhage at the sites of metastases containing high-volume of choriocarcinomatous elements with significant elevation of serum level of β-hCG levels in patients diagnosed with testicular carcinoma is known as choriocarcinoma syndrome.

Keywords: hemorrhage, metastasis, raised β-hCG, testicular malignancy.

Testicular neoplasms comprise the most common solid malignancy affecting men between the ages of 15 and 35, but they only represent almost 1% of all solid tumors in males. Approximately two to three new cases per 100,000 males are reported in the United States each year, and 95 percent of all primary testicular tumors are germ cell tumors.[1] This rare tumor can be complicated by a very rare, life threatening complication which is choriocarcinoma syndrome with only few cases reported worldwide. Choriocarcinoma syndrome entails hemorrhage from metastatic sites of choriocarcinoma associated with a significant rise of beta-human chorionic

gonadotropin (β-HCG).[2] In this report, we present a case of choriocarcinoma syndrome in a 25 years gentleman, to show the importance of considering this syndrome while dealing with patients who have advanced testicular tumors with high β-HCG, as this syndrome is life-threatening and needs urgent treatment.

Case Report

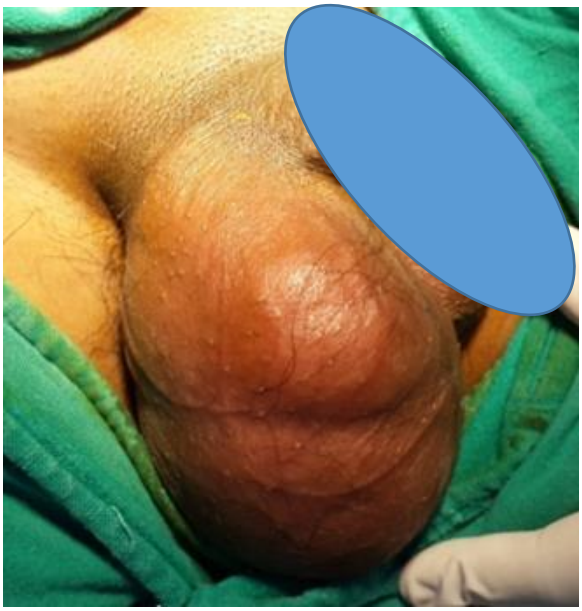

Figure 1: Plysical appearance of right testicular tumor

A 25-year-old gentleman presented to the urology outpatient department with the diagnosis of painless, non-progressive right sided scrotal swelling for 9 months. There was no history of local trauma. No urinary symptoms. No history of weight loss, jaundice or fever. On examination, he was well built gentleman. Vitals signs were all within normal limits. No anemia, jaundice or lymphadenopathy. There was 8x8x6 cms bilobulated hard mass over the right scrotal region which was non-tender, smooth and non-transilluminant. Right testis could not be palpated separately. Left sided scrotum normal grossly **(Figure 1)**. Systemic evaluation were normal.

His hematological parameters showed slight leukocytosis (11,700), raised ESR 30 mm/Hr with slightly below par hemoglobin levels of 11.9 gm%. Liver function test and kidney function test parameters were within normal limits. He had normal bleeding time and clotting time but prothrombin time INR was mildly increased to 1.15. USG scrotum **(Figure 2 A)** showed a 9x8x7 cms vascular solid mass in the right scrotum replacing the whole of the right testis, most likely to be malignancy. Left testis appeared normal. Also, the USG abdomen and pelvis showed

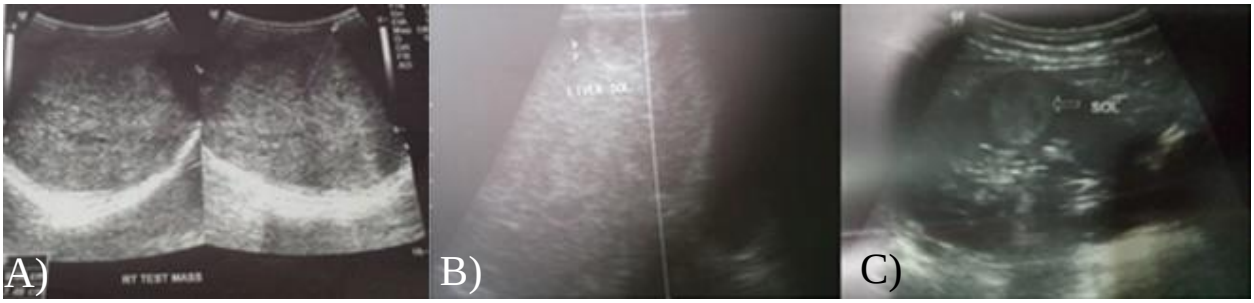

Figure 2: A) USG appearance of testicular tumor, B) USG Showing liver metastasis, C) USG showing renal metastasis

well defined echogenic lesion of heterogeneous echotexture at the medial aspect of mid pole of spleen, liver **(Figure 2 B)** and mid pole of right kidney **(Figure 2 C)** most likely metastasis.

Figure 3: Right testis specimen

Testicular malignancy tumor markers showed raised serum lactate dehydrogenase (LDH) levels of 1951 IU/L, raised alpha-fetoprotein (AFP) levels of 1590 ng/ml and markedly raised β-hCG levels of 5,02,110 IU/L. There were also two cannon ball shadows over the bilateral lower lung zones suggestive of malignancy. Thereafter, radical right orchidectomy via high inguinal approach was done **(Figure 3)**.

Histopathological report showed mixed germ cell tumor (80% seminomatous and 20% choriocarcinomatous component). Spermatic cord and tunica vaginalis free of tumor extension with no lymphovascular invasion.

On immediate post-operative period, patient developed sudden abdominal distension and tenderness. Hb level decreased upto 4.5 gm%. Patient was transferred to high dependency unit and required multiple units of blood transfusion. On 3rd post-operative day, CECT head, chest, abdomen and pelvis showed multiple metastases at the left occipital region, bilateral lower lung zones, right kidney, spleen and liver respectively. No obvious regional lymphadenopathies.

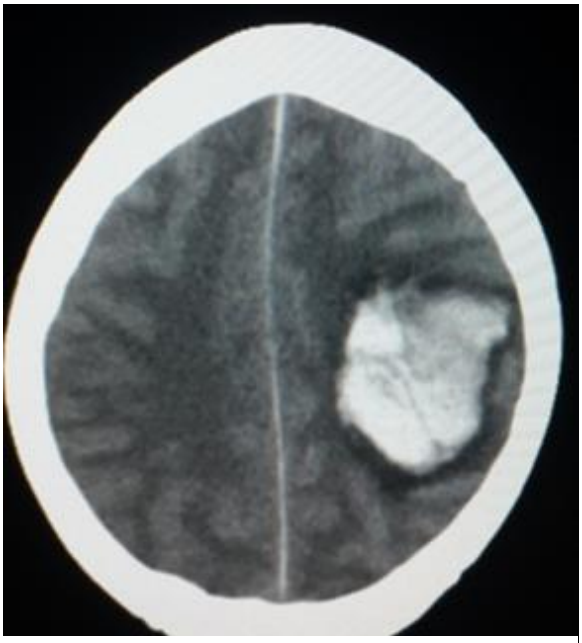

Figure 4: Intracerebral hemorrhage

Most significantly patient had hemorrhagic regions over metatstatic region of right kidney. This led to the diagnosis of pT1N0M1 mixed germ cell tumor (Stage III) choriocarcinoma syndrome. Patient was started on Etiposide (100mg) + Paclitaxel (2mg) as per the regimen discussed with the medical oncologist. However on the 6th day

of chemocycle there was sudden loss of consciousness. No SOB, hemoptysis or abdominal distension. CECT head showed large hemorrhagic foci over the left lacunar region **(Figure 4)**. Due to poor prognostic outcome further surgical intervention was refused and patient eventually expired after two months at outer center.

Discussion

We present the clinical course of a patient with a metastatic testicular cancer which was complicated with choriocarcinoma syndrome as he developed bleeding in the renal and brain metastases leading to his death. Choriocarcinoma syndrome was first described by Logothetis in 1984. It was characterized by hemorrhage at the site of metastases containing high-volume choriocarcinomatous elements with significant elevation of serum levels of β-hCG.[2]

The pathogenesis of choriocarcinoma syndrome is unknown. It may be related to tumor invasion of the small blood vessels.[3] It occurs in two different clinical settings, either few hours after initiation of combined chemotherapy which is more common or much less likely spontaneously in advanced disease without relation to treatment as in our patient.[4] Acute hemorrhage in the pulmonary metastasis is the typical presentation of choriocarcinoma syndrome; however, hemorrhage at any site of metastasis can develop.[2]

In our patient, there was spontaneous hemorrhage mainly in renal and intra-cerebral region although there were metastasis in the lungs as well. Apart from this hyperthyroidism can be paraneoplastic presentation of choriocarcinoma syndrome. The mechanism of the hyperthyroidism is probably due to the ability of HCG to stimulate the TSH receptors, as it has an identical alpha subunit to that of the TSH.[5] Till date no guidelines or treatment protocols can clearly define the management of choriocarcinoma syndrome. M. Tatokoro and colleagues have reported the only surviving case of choriocarcinoma syndrome where the patient was salvaged after emergency lower lobectomy for hemothorax due to rupture of pulmonary metastases. Hence, early recognition and urgent treatment which is usually multimodal consisting of medical treatment, usually in the intensive care unit, to stabilize the patient and surgical intervention when appropriate.[6]

Conclusion

Choriocarcinoma syndrome should be considered when dealing with patients diagnosed with germ cell type testicular tumor with markedly raised β-hCG levels and with metastasis. Urgent intervention may be only option. That being said, prognosis is always grave.

References

1. J. M. Stephen, M. Papadakis, and M. W. Rabow, Current Medical Diagnosis and

Treatment. 50th ed. San Francisco, Calif, USA: McGraw-Hill Medical, 2011.

2. C. J. Logothetis, M. L. Samuels, and D. E. Selig, Cyclic chemotherapy with cyclophosphamide, doxorubicin, and cisplatin plus vinblastine and bleomycin in advanced germinal tumors. Results with 100 patients, The American Journal of Medicine. 1986;81:219–27.
3. M. Shintaku, M. H. Hwang, and R. Amitani, Primary choriocarcinoma of the long manifesting as diffuse alveolar hemorrhage, Archives of Pathology and Laboratory Medicine. 2006;130:540–3.
4. R. J. Motzer and G. J. Bosl, Hemorrhage: a complication of metastatic testicular choriocarcinoma. Urology. 1987;30:119–22.
5. S. F. Oosting, E. C. de Haas, T. P. Links, Prevalence of paraneoplastic hyperthyroidism in patients with metastatic non-seminomatous germ-cell tumors, Annals of Oncology, 2010;21:104-8.
6. M. Tatokoro, S. Kawakami, M. Sakura, T. Kobayashi, K. Kihara, and H. Akamatsu, Successful management of life-threatening choriocarcinoma syndrome with rupture of pulmonary metastatic foci causing hemorrhagic shock, International Journal of Urology. 2008;15:263–648.