Retroperitoneal Endodermal Sinus Tumor Patient with Palliative Care Needs

Surbhi Kashyap

Windsor University School of Medicine, St. Kitts, West Indies

Address for correspondence: Dr. Surbhi Kashyap; E-mail: kashyapsurbhi@yahoo.com

ABSTRACT

This article is a case reflection of a personal encounter on the palliative care treatment required after the removal of a complicated case of a primary extra-gonadal retro-peritoneal endodermal sinus tumor (yolk sac tumor). This reflection is from the perspective of a recently graduated MD student who spent one month with an Indian pain management and palliative care team at the Institute Rotary Cancer Hospital (IRCH), All India Institute of Medical Sciences (AIIMS), New Delhi

Key words: Health literacy, Neuropathic pain, Pain management, Palliative care, Retroperitoneal endodermal sinus tumor, Yolk sac tumor

INTRODUCTION

Yolk sac tumors (YST) are nonseminomas germ cell tumors that originally arise from testes or ovaries. Primary extragonadal germ cell tumors (EGCTs) are extremely rare, which account for 1–4% of all germ cell tumors. EGCT's are typically found in the midline.[1,2] The most common extragonadal sites are sacrococcygeal, mediastinal, intracranial, and retroperitoneal.[1,2] In this article, we will be discussing a rare case of a primary extragonadal retroperitoneal YST with spinal cord metastasis, treated with surgery, and pain medications but not chemotherapy.

CASE REPORT

A 12-year-old male child with a 3-month history of bilateral, constant, radiating lower limb, and back pain was brought to Institute Rotary Cancer Hospital (IRCH) by his father. The patient described the pain as an aching, throbbing, stabbing pain, and rated it a 9/10 on the Verbal Analogue Scale (VAS). He stated that he had not been able to walk or do any of his regular physical activities for a month. He had associated weakness and paresthesias in his legs bilaterally. He was unable to sleep at night for the past 2 weeks, due to back pain that was elicited by lying down. The patient also had associated constipation and urinary retention for which he had to be catheterized.

When the patient came to the pain clinic, he was held in his father's arms, as the patient was paralyzed from the waist down. On physical examination, the patient appeared to be in severe pain. Deep tendon reflexes were not present in bilateral lower limbs. Muscle strength was 2/5 on the Oxford Scale (muscle strength grading scale) in bilateral limbs (hip/knee/ankle). Sensation to pinprick and soft touch were diminished over the gluteal

Access this article online

Quick Response Code:

Website: www.jpalliativecare.com

DOI: 10.4103/0973-1075.173955

How to cite this article: Kashyap S. Retroperitoneal endodermal sinus tumor patient with palliative care needs. Indian J Palliat Care 2016;22:97-100.

regions and legs. Upon palpation of the patient's legs, he felt electricity running through his legs. The patient denied any other symptoms.

In March 2015, the patient had sudden onset of excruciating left lower quadrant abdominal pain. In April 2015, he underwent an exploratory laparotomy and excision of a growth between the spleen and renal space. Pathology reports suggested the growth was a CD99 and vimentin-positive YST. Further tests were conducted such as an ultrasound to see if there was any involvement of the testes. Results showed no gonadal involvement.

The patient's father stated that the patient was attending school, running, and was pain-free until 15 days postoperatively. The doctors at his local hospital were unable to find the etiology of his postoperative pain. The patient was referred to a different hospital for consultation. A CT scan was done that showed a heterogeneous mass in the left suprarenal region, suggestive of a residual mass that was infiltrating the posterior abdominal wall and muscle. A whole abdomen contrast-enhanced computed tomography (nonionic CECT) revealed a devascularized left kidney possibly due to left renal artery injury. After various inconclusive testing, the patient was referred to IRCH (AIIMS.)

Management

At the time of the patient's presentation at IRCH his condition had progressively worsened. He had developed bilateral paralysis, severe lower lumbar back pain, and bilateral leg pain. Many investigations were initiated which included complete blood count (CBC), liver function tests, complete metabolic panel, blood test checking for thyroid-stimulating hormone (TSH), beta-human chorionic gonadotropin (β-HCG), and alpha-fetoprotein (AFP) levels, comparison CECT scan, ultrasound of neck to check for lymph node involvement.

CBC showed hemoglobin was 10.4 (normal range: 13.5–17.5 g/dl for males), thin layer chromatography was 24.8 (normal range: 4000–11,000). Liver function tests showed that bilirubin levels were below the normal range at 0.2 mg% (normal range: 0.8–1 mg%), TSH and β-HCG were within normal limits, but AFP 3 was 0.52 ng/ml (normal range: 0.89–8.78 ng/ml). Ultrasound of the neck showed no sign of metastasis to lymph nodes or glands. The CECT scan was unchanged compared to the previous scan. The mass seen previously was causing erosion of the left 12th rib with extension into neural foramina with suspicious intraspinal extension. No metastatic lung nodules were visible.

Based on the patient's history, physical exam and investigations, the diagnosis of primary retroperitoneal extragonadal YST extending into the neural foramina of the spinal cord at L1/L2 was made. The metastasis was causing compression of the spinal cord producing his paralysis. For management of this patient the medical oncologist, surgical oncologist, and radiologist were consulted. Due to the extent of cancer, further surgery was no longer an option.[3] Although the paralysis was not reversible, the pain, with the help of proper pain management, and palliative care team was treatable.[4]

While interacting with the patient and his father, I could see that the patient's father was distraught about his son's condition. Medical treatments for the patient had taken a financial toll on the family. The patient's father had to sell off his land where he was farming in Patna, Bihar in order to pay for proper treatments. He expressed to me that he was willing to sell his house too if it meant we could make his son walk again. The patient's mother became severely depressed and threatened to commit suicide if her son's health did not improve. Despite explaining that at this stage only the pain of the patient could be managed and that the paralysis was permanent, the father was in denial. He kept saying, "Please make my son walk again, that is, all I want." Family bonds are reasonably vulnerable during the time of distress, and the entire palliative care team provided a positive and supportive environment so that they could communicate their needs most effectively.

The patient was initially started on infusion morphine, which was dose titrated appropriately and later converted to tablet morphine.[4,5] Dexamethasone was prescribed to reduce the swelling caused by the compression of the spinal cord.[6] He was given gabapentin for the neuropathic pain, which was converted, to tablet pregabalin. Studies suggest pregabalin provides equivalent efficacy to gabapentin, however, at much lower doses.[7,8] Because lower dosages can be used to treat neuropathic pain, it is likely that pregabalin will be associated with fewer dose-related adverse events such as dizziness, chest pain, difficulty breathing, and constipation.[7-9] Pregabalin requires lower dosages, has a much higher bioavailability (90% vs. 33–66% of gabapentin) and is rapidly absorbed (peak: 1 h).[4,7,8]

Morphine reduced the pain from 9/10 to 4/10 on the VAS.[4,5] To further relieve the patient's pain, we discussed chemotherapy and attempting a nerve block.[10] The

father cited apprehensions for both of these treatment options. The patient's father was explained the positive impact the procedure would have on relieving his son's pain, and that his son would be able to sleep peacefully and be pain-free. He was told giving chemotherapy would not have reversed the neurological deficit but would have definitely reduced the pain by treating the cause. The father had seen many unfortunate situations about what can happen after chemotherapy is given and explained to us how he knew some people that had died in his village after chemotherapy was given. He was further explained all the benefits chemotherapy would have on not only the pain the patient was experiencing but also in slowing down the progression of metastasis. All his questions were addressed, and he was told that the situation in which other people might have received chemotherapy might be different than his son's. When explained how attempting a nerve block could be beneficial, the father again refused and said he heard it would permanently damage his son's nerves. The whole palliative care team made many efforts to make the patient's father understand exactly how these treatments would be given as well as how all the side effects would be managed for both the nerve block and chemotherapy, but the father was convinced that these will only cause harm to his son based on what he had seen and what people told him about these medical advances in his village. Even after multiple attempts of ensuring the father that this would be the next best option for his son's pain, consent was not given, so these treatments were not given.[11] One of the major components of palliative care is respect for patient's autonomy.[11] Due to lack of health literacy there are many proven, working treatments that are simply not done.[12]

The patient's pain was managed based on the World health Organizations Cancer pain protocol (WHO ladder).[13,14] This included the analgesics tablet Morphine (P) 10 mg three tablets 4 hourly, tablet paracetamol (acetaminophen) 500 mg one tablet QID and tablet pregabalin 75 mg HS × 6 weeks.[8,13] Anti-emetics were given to avoid nausea and vomiting, the patient was experiencing.[13] For GI ulcer prophylaxis tablet pantoprazole 40 mg once daily was prescribed. Tablet dexamethasone 4 mg TDS × 7 days was also given and later changed to 2 mg TDS × 7 days.[4,7,8,13] After monitoring the patient's pain and bringing it down to a 3/10 on the VAS, the patient was counseled and sent to a nonprofit organization called Cankids (Cankids is a registered charitable national society working to make a change for childhood cancer in India.) There, he would receive further pain management treatments and psychosocial support through counseling for him and his family.

DISCUSSION

Endodermal sinus tumor is the most common pure malignant germ cell tumor in young children with poor prognosis.[15] An exclusively retroperitoneal or abdominal location for an endodermal sinus tumor is uncommon, comprising less than 5% of all EGCTs.[15,16] Most YSTs will require surgery and chemotherapy, regardless of stage or presence of metastasis, because of the aggressive nature, and recurrence of the disease.[17,18] Complete resection of retroperitoneal tumors is necessary for successful treatment.[16,19]

AFP is an important tumor marker for YSTs. An increased serum AFP level is typically observed in patients presenting with an YST. Serum AFP levels decrease rapidly following tumor resection, however, the levels increase during tumor recurrence or metastasis.[16,20] It has also been noted that sometimes AFP levels may decrease or remain normal 5–7 weeks after surgery.[5] The sensitivity of AFP for nonseminomas germ cell tumors is 50–60%, the sensitivity of β-HCG is 30–60%.[5,20] Therefore, the combined use of AFP and β-HCG is more effective than either marker alone. In our case, the patient's β-HCG was within normal limits and AFP was decreased. Only 75% of patients actually show an increase in β-HCG and AFP, our patient fell into the 25% of patients with a decrease AFP level.[5,20]

Insufficient pain control can be very debilitating in metastasizing childhood cancers and can interfere with physical rehabilitation, mobility, and proper nutrition.[21] Chemotherapy, nerve blocks, radiofrequency ablations, neurolytic destructions, as well as many other procedures are less invasive than surgery but can improve the pain drastically when medications do not suffice.[17] The lack of health literacy in developing countries is causing many patients to receive inadequate treatments. Health literacy is a combination of reading and comprehending along with decision-making skills, and the ability to apply these skills to various health-related situations.[12] The development of health literacy should be considered as a way of improving overall health through proper communication.[12] If consent was provided more appropriate pain management directed procedures could have been performed to bring the pain from 4/10 to possibly 2/10 or lower.

CONCLUSION

In the 21st century, medical advancements have left us with little to none untreatable conditions. One of the few problems that remain is the widespread lack of health literacy. If we could increase the global health literacy, we

would be able to help patients with treatable conditions. This case has shown me that even treatable conditions and in-depth explanations to patients cannot overcome the deep-rooted lack of health literacy (via fear of treatment modalities and possible side-effects) that interferes with proven treatments of various illnesses.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Göbel U, Schneider DT, Calaminus G, Haas RJ, Schmidt P, Harms D. Germ-cell tumors in childhood and adolescence. GPOH MAKEI and the MAHO study groups. Ann Oncol 2000;11:263-71.
2. Hsu YJ, Pai L, Chen YC, Ho CL, Kao WY, Chao TY. Extragonadal germ cell tumors in Taiwan: An analysis of treatment results of 59 patients. Cancer 2002;95:766-74.
3. Leleszi JP, Lewandowski JG. Pain management in end-of-life care. J Am Osteopath Assoc 2005;105 3 Suppl 1:S6-11.
4. Hanks GW, Conno F, Cherny N, Hanna M, Kalso E, McQuay HJ, *et al*. Morphine and alternative opioids in cancer pain: The EAPC recommendations. Br J Cancer 2001;84:587-93.
5. Talerman A, Haije WG, Baggerman L. Serum alphafetoprotein (AFP) in diagnosis and management of endodermal sinus (yolk sac) tumor and mixed germ cell tumor of the ovary. Cancer 1978;41:272-8.
6. Raj, Vishwa S, LaTanya Lofton. Rehabilitation and Treatment of Spinal Cord Tumors. The Journal of Spinal Cord Medicine 2013;36:4-11.
7. Bockbrader HN, Wesche D, Miller R, Chapel S, Janiczek N, Burger P. Acomparison of the pharmacokinetics and pharmacodynamics of pregabalin and gabapentin. Clin Pharmacokinet 2010;49:661-9.
8. Freynhagen R, Strojek K, Griesing T, Whalen E, Balkenohl M. Efficacy of pregabalin in neuropathic pain evaluated in a 12-week, randomised, double-blind, multicentre, placebo-controlled trial of flexible- and fixed-dose regimens. Pain 2005;115:254-63.
9. Moore, R Andrew *et al*. Pregabalin for Acute and Chronic Pain in Adults." The Cochrane database of systematic reviews 2009;3:CD007076.
10. Abrahm JL, Banffy MB, Harris MB. Spinal cord compression in patients with advanced metastatic cancer: "All I care about is walking and living my life". JAMA 2008;299:937-46.
11. Devakaran E. Ethical Issues of Palliative Care. Thrissur: Essentials of Palliative Care; Institute of Palliative Care, 2015. p. 39.
12. Ratzan SC. Health literacy: Communication for the public good. Health Promot Int 2001;16:207-14.
13. Ripamonti CI, Bandieri E, Roila F; ESMO Guidelines Working Group. Management of cancer pain: ESMO Clinical Practice Guidelines. Ann Oncol 2011;22 Suppl 6:vi69-77.
14. Hooke C, Hellsten MB, Stutzer C, Forte K. Pain management for the child with cancer in end-of-life care: APON position paper. J Pediatr Oncol Nurs 2002;19:43-7.
15. Murat E, Dagdemir A, Bilgici MC, Süllü Y. Primary yolk sac tumor of the retroperitoneal region. Contemp Oncol (Pozn) 2013;17:530-2.
16. Guo YL, Zhang YL, Zhu JQ. Primary yolk sac tumor of the retroperitoneum: A case report and review of the literature. Oncol Lett 2014;8:556-60.
17. Nersesyan H, Slavin KV. Current aproach to cancer pain management: Availability and implications of different treatment options. Ther Clin Risk Manag 2007;3:381-400.
18. Chambers WA. Nerve blocks in palliative care. Br J Anaesth 2008;101:95-100.
19. DiPerna CA, Bowdish ME, Weaver FA, Bremner RM, Jabbour N, Skinner D, *et al*. Concomitant vascular procedures for malignancies with vascular invasion. Arch Surg 2002;137:901-6.
20. Talerman A, Haije WG, Baggerman L. Serum alphafetoprotein (AFP) in patients with germ cell tumors of the gonads and extragonadal sites: Correlation between endodermal sinus (yolk sac) tumor and raised serum AFP. Cancer 1980;46:380-5.
21. Cherny NI. The management of cancer pain. CA Cancer J Clin 2000;50:70-116.