Sri Lanka Journal of Medicine SLJM

Case report

Citation: Arudchelvam J, Kumari LEDI, Manathunga R, et al. A Rare Occurrence of Adrenal Neuroblastoma in an Adult: Case Report. SLJM. 2026, 35 (2): 55- 58
DOI: https://doi.org/10.4038/sljm.v35i2.745

A Rare Occurrence of Adrenal Neuroblastoma in an Adult: Case Report

J Arudchelvam[1,2], L E D I Kumari[2], R Manathunga[3], RSP Premarathna[3]

[1]Department of Surgery, Faculty of Medicine, University of Colombo, Sri Lanka

[2]National Hospital of Sri Lanka, Colombo, Sri Lanka

[3]Apeksha Hospital Maharagama, Sri Lanka.

Correspondence:

Arudchelvam J

E mail: joelaru@srg.cmb.ac.lk

https://orcid.org/0000-0002-4371-4527

ABSTRACT

Adrenal neuroblastoma (ANB) originates from neural crest cells in the adrenal medulla and is rare in adults, with fewer than 10% of cases occurring after age 14 and with a prevalence of about 0.12 per million. Clinical presentations are nonspecific, often including abdominal pain or mass, leading to delayed diagnosis. Imaging typically shows a heterogeneous adrenal mass. Due to its rarity, adult cases are treated using paediatric protocols. ANB is highly aggressive and carries a poor prognosis in adults. This report describes a 17-year-old male with a large ANB, successfully treated with radical surgical resection and chemotherapy, followed by an uneventful recovery.

Keywords: adrenal neuroblastoma; Adult; Imaging in adrenal neuroblastoma

INTRODUCTION

Adrenal neuroblastoma (ANB) arises from the primordial neural crest cells in the adrenal medulla. It is rare in adults and only few cases are reported (about 151 cases in adolescents reported so far) [1] . Out of all ANB, less than 10% occur in individuals more than 14 years of age, with an incidence of 0.12 cases per million [2], [3].

The clinical presentation of ANB is nonspecific with abdominal mass and pain, therefore the diagnosis is often delayed [3]. On imaging the ANB appears as a heterogeneous mass lesion arising in relation to the adrenal gland.

Due to its rarity in adults, the ANB in adults are managed with paediatric management protocols. The ANB is an aggressive tumour and is associated with very poor prognosis in adults [4]. This case report describes a 17-year-old male who presented with a large ANB (16.7cm * 15.4 cm). He was managed with neoadjuvant chemotherapy followed by radical excision (with en bloc nephrectomy, distal pancreatectomy and splenectomy). The patient made an uneventful recovery.

CASE REPORT

A 17-year-old male presented with a history of epigastric pain for four months duration. On examination, there was a mass in the epigastric and left hypochondriac region. An ultrasound scan (USS) of the abdomen showed a large heterogeneous mass with cystic and solid areas arising from the left suprarenal area. A contrast enhanced computed tomography (CT) of the chest and abdomen was done. CT demonstrated a large mass arising from the left suprarenal gland. The mass was displacing the aorta and the

inferior venal cava (IVC) to the right side (Figure 1). The mass was encasing the aorta (about 180 degrees). In addition, the coeliac axis, superior mesenteric artery and the left renal vessels were displaced. The tissue planes between the tumour and the aorta were clearly defined, except for a small area near the supracoeliac aorta where the planes were not well defined. The left kidney was displaced inferiorly. There was no clear soft tissue plane demonstrated between the kidney and the mass. The pancreatic body and tail did not have a clear tissue plane between them and the mass. There was no evidence of distant metastasis. The urine Vanillylmandelic Acid (VMA) level and the 24-hour urinary metanephrine excretion (158.9 µg/ 24-hour (normal < 350) levels were normal.

Since the plane between the tumour and the supracoeliac aorta was not well defined, a magnetic resonance imaging (MRI) of the abdomen was done. MRI demonstrated a 16.7cm * 15.4 cm, heterogeneous mass lesion with displacement of the aorta and the IVC to the right side (Figure 2). The coeliac axis was also displaced by the mass. However, the tissue plane between the mass and the major vessels was well defined.

Depending on the radiological and clinical features, a neuroblastoma of the adrenal gland was suspected. Following a multidisciplinary discussion with the oncologist, because of the high-risk features, a pre-operative biopsy and neoadjuvant chemotherapy was planned. The biopsy confirmed the ANB (neuron specific enolase (NSE) - positive). The patient was given three cycles of cyclophosphamide, doxorubicin, and vincristine (CAV), followed by two cycles of cisplatin and etoposide (PE).

Following the neoadjuvant chemotherapy, a radical excision of the adrenal gland tumour, the left kidney, the distal pancreas and the spleen was planned. A midline laparotomy with a transverse extension to the left, starting from two inches above the umbilicus was done. The descending colon, the splenic flexure, and the transverse colon were mobilized medially. The left colic artery (LCA) was found to be running through the tumour. Therefore, the LCA was ligated and divided. Following the ligation of the LCA, the colon remained viable. Therefore, colectomy was not performed. The stomach was mobilised away from the spleen by dividing the gastro splenic ligament. The tumour, spleen, distal pancreas, and the left kidney were mobilized with the surrounding retroperitoneal fat. The tumour was carefully dissected away from the major vessels. Large suprarenal arteries supplying the tumour were ligated and divided. The pancreas neck was divided. That tumour was removed en bloc with the distal pancreas, spleen and the left kidney (Figure 3). The patient made an uneventful recovery.

The histology demonstrated a tumour with solid and necrotic areas. The tumour was covered by a thick capsule. Microscopically the tumour consisted of neuroblasts and ganglion cells, in varying stages of differentiation in a background of loosely arranged schwannoma stroma. The features were consistent with a poorly differentiated neuroblastoma with invasion into the pancreas. One of the three lymph nodes was positive for tumour cells. There was no invasion into the spleen or the kidney. The patient was referred to the oncologist for further management.

DISCUSSION

Adrenal neuroblastoma is common in children. About 90.0% of the neuroblastomas occur in children less than 5 years of age, and it is rare in adults. ANB in adults occur in individuals between 18 to 60 years with a male predominance (about 60.0% of the case are males). In a review of adults with neuroblastoma, the mean age was 41.6 years [1,3].

The commonest sites for the adult neuroblastoma are the adrenal glands (50.0%), followed by retroperitoneal structures (17.0%). Other sites of adult neuroblastoma include neck, chest and pelvis [5]. The ANB is associated with very poor prognosis in adults due to its aggressive growth and the metastasis [4]. About 50.0% of the patients have liver, lymph node and bone marrow metastasis at the time of presentation [6].

The patients with neuroblastoma most commonly present with abdominal mass and abdominal pain. In a review of 127 studies, the mean tumour size at presentation was 9.95 cm (4.5 – 34). In the same review, 56.0% of the tumours were more than 10.0 cm in size. In the above reported case, the size was 16.7cm. These findings indicate that the patients do not become symptomatic until the mass is of a large size [1]. Other patients present with features of metastasis and endocrine manifestations (e.g. tachycardia, hypertension and sweating due to the catecholamine secretion [7,8]. Other presentations include opsomyoclonus i.e. a combination of ataxia, irregular movements and rapid eye movements. Another described presentation is the Kerner-Morrison syndrome, due to the secretion of vasoactive intestinal peptide (VIP) that leads to watery diarrhoea.

Imaging is essential in diagnosis, staging and differentiating the neuroblastoma from the other

retroperitoneal tumours and the renal malignancies. USS can identify the organ of origin. USS also demonstrates the cystic, solid components and the intra mass calcifications. However, USS is not a good investigation to demonstrate the extent of the vascular involvement and the local invasion.

The CT scan shows a globular cystic and solid tumour with heterogeneous density. CT also demonstrates the necrotic areas and the intra tumour calcification. The CT scan is a good investigation to demonstrate the extent of local invasion and the vascular encasement. However, The CT scan does not demonstrate the tissue planes well, especially when the surrounding tissues are compressed by the tumour [9].

The magnetic resonance imaging (MRI) demonstrates a heterogeneous, hyper intense tumour on T2 and hypo intense tumour in T1 weighted images. It also demonstrates the tumour with solid and cystic areas, calcification and degeneration. The MRI is a better investigation to demonstrate the soft tissue planes, especially in case of suspected vascular wall involvement [10]. MRI is also better in detection of bone marrow and liver secondaries and the spinal cord involvement [6].

The ANB is staged according to the International Neuroblastoma Risk Group (INRG) staging system. This system depends on image-based risk factors and provides risk stratification. This system predicts the surgical outcome and guides the management.

Due to its rarity in adults, the ANB in adults are managed with paediatric protocols [4].The management of neuroblastoma includes a combination of surgical excision, chemotherapy and radiotherapy. Surgery alone is offered for low-risk tumours.

Complete surgical excision is the essential for cure and prevention of recurrence. However, complete surgical resection may be difficult due to the size of the ANB and the invasion into the surrounding tissues. In a review of 127 articles, complete reception (R0) was achieved only in 72.0% of the cases. One study reported that the five-year survival with complete resection was 41.2%, while in patients with incomplete resection the survival was only 18.7% [11]. The major reasons given in the literature for the incomplete resection are vascular encasement and the presence of extensive lymph node metastasis.

The neoadjuvant chemotherapy is given for high risk ANB as in the case described above. High risk features include, large tumours and the presence of extensive metastasis. Neoadjuvant chemotherapy is known to reduce the tumour size. However, in the case described above, there was no significant reduction of the tumour size following the neo adjuvant chemotherapy. Cisplatin based drugs (cisplatin/etoposide or carboplatin/doxorubicin) are commonly used as neoadjuvant agents. In the above-described case, doxorubicin and cisplatin were used in combination with other drugs.

Adjuvant chemotherapy is given with a combination of drugs, in cases with advanced primary and in patients with metastatic ANB. The drugs used for the adjuvant chemotherapy are cyclophosphamide, adriamycin, carboplatin, cisplatin, and etoposide. Radiotherapy is also used in high-risk cases and in cases with high chances of recurrence i.e. large-size tumours and cases with positive margins. Studies have reported that the adjuvant chemotherapy and radiotherapy following surgery Improves the survival from 28.5% to 42.8% [11].

The ANB is associated with very poor prognosis in adults due to its aggressive growth and the metastasis [4]. Complete excision followed by adjuvant therapy is known to reduce the recurrence and to improve survival. Therefore, early diagnosis, neoadjuvant and adjuvant therapy is essential to achieve long term survival. This case report presents the surgical aspects and the immediate outcome of the ANB. The long-term follow-up of this case and reporting such cases is important to add to the knowledge of this rare condition in adults.

CONCLUSION

Therefore, adrenal neuroblastoma in adults is rare, which poses significant challenges in diagnosis and management. Due to its rarity in adults, standardised treatment protocols are not well established. However, complete surgical excision of the tumour remains the cornerstone of treatment, as it is shown to reduce the risk of recurrence. Adjuvant therapies, such as chemotherapy and radiotherapy, are also shown to further improve the survival. This case report focuses on the surgical aspects and the immediate postoperative outcome of an adult patient with adrenal neuroblastoma, demonstrating the feasibility and the importance of complete tumour removal. Given the scarcity of adult cases, long-term follow-up is essential to monitor for potential recurrence and to assess the effectiveness of combined treatment strategies over time. Reporting such rare cases contributes significantly to the limited existing literature.

Author declaration

Acknowledgement:
None.

Authors' contributions:
JA : conceptualisation, wrote the manuscript, images, done the literature survey, involved in the management of the patient and reviewed; L E D I K : involved in the management of the patient and performed data collection; RM : involved in the management of the patient and performed data collection; RSPP : involved in the management of the patient and performed data collection. All authors reviewed and approved the final manuscript.

Conflicts of interest:
The authors declare that they have no conflicts of interest.

Funding statement:
This research received no external funding.

Ethics statement:
All patient data were anonymized and managed strictly to ensure confidentiality. Written informed consent was obtained from the patient for the publication.

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