

Poland syndrome and renal cell cancer

Ruken Yuksekkaya, Fatih Celikyay, Caglar Deniz, Berat Acu

Department of Radiology, Gaziosmanpasa University School of Medicine, Gaziosmanpasa Universitesi Tip Fakultesi Hastanesi Radyoloji Anabilim Dalı, Tokat, Turkey

ABSTRACT

Poland's syndrome is a rare congenital anomaly characterized by chest wall and upper limb abnormalities. Some lymphoreticular and solid malignancies have been reported with this syndrome. Renal cell carcinoma associated with Poland's syndrome has not been described previously. We herein describe the first case of a Poland's syndrome associated with renal cell carcinoma diagnosed incidentally after a road accident. All the cases with Poland's syndrome should be aware of oncologic attention by the radiologists and clinicians.

Key words: Rare cancer, poland's syndrome, renal cell carcinoma

INTRODUCTION

Poland's syndrome is a rare congenital anomaly consists of absence or hypoplasia of the pectoralis major muscles, malformation of the upper limbs, breasts and nipples, ribs and costal cartilages, chest wall subcutaneous tissues. This syndrome may be associated with hypoplasia of the subclavian artery or its branches which may lead to different developmental changes.^[1] Cases of Poland's syndrome associated with lymphoreticular malignancies,^[2] and solid tumors as leiomyosarcoma,^[3] neuroblastoma,^[4] Wilm's tumor,^[5] breast cancer,^[6] lung cancer,^[7] and gastric cancer,^[8] were reported. These may confirm the relationship between developmental defects and tumors and require oncologic attention.

To our knowledge, we herein describe the first case of a Poland's syndrome associated with renal cell carcinoma diagnosed incidentally after a road accident. The aim of this article is to evaluate the multidetector computed tomography (MDCT) findings of Poland's syndrome and draw the attention of the radiologists and clinicians in these cases may have increased risk of malignancies.

CASE REPORT

A 51-year-old unconscious patient after a road accident came to our department for the evaluation of the thorax with chest radiography and MDCT. On supine anteroposterior chest radiography left hemithorax was slightly hiperlulent. The MDCT examinations were performed on a eight channel MDCT system (GE Medical Systems, Milwaukee, WI, USA). MDCT revealed pleural effusion, compression atelectasis, minimally displaced and fragmented fractures of the ribs II to V on the right hemithorax. On the left hemithorax, the pectoralis major and minor, subscapularis, supraspinatus, infraspinatus, serratus anterior, teres major and minor muscles were absent. Erector spina and trapezius muscles were hypoplastic [Figure 1]. These findings confirmed the presence of Poland's syndrome. In the abdominal sections, a lobulated and heterogenously enhancing mass with cystic areas at the right kidney was observed [Figure 2]. The mass was extended to the perirenal fat planes and renal sinus. The imaging appearance of the renal mass suggested renal cell carcinoma. The patient underwent radical nephrectomy at another institution. Pathological examination revealed renal cell carcinoma.

DISCUSSION

Poland's syndrome was first described by Alfred Poland, in 1871. The incidence of the syndrome is between 1:7000

Access this article online

Quick Response Code:



Website:

www.cci-j-online.org

DOI:

10.4103/2278-0513.99570

Address for correspondence: Ruken Yuksekkaya, Department of Radiology, Gaziosmanpasa University School of Medicine, Gaziosmanpasa Universitesi Tip Fakultesi Hastanesi Radyoloji Anabilim Dalı, Tokat, Turkey. E-mail: rukenyuksekkaya@yahoo.com



Figure 1: Axial MDCT image shows the absence of pectoralis major and minor, subscapularis, supraspinatus, infraspinatus, serratus anterior, teres major and minor muscles and hypoplasia of erector spina, trapezius muscles

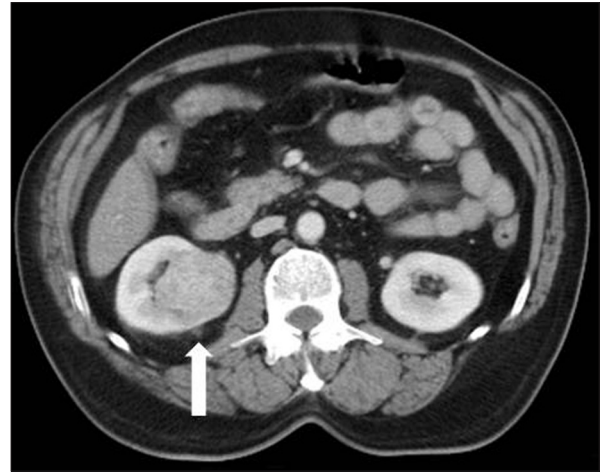


Figure 2: Axial MDCT image reveals a lobulated and heterogeneously enhancing mass with invasion of the renal sinus and perirenal fat planes at the right kidney

and 1:100,000. It is a nongenetic, congenital disorder. The features of the syndrome show differences. It is mostly unilateral. The major components of the syndrome are the absence of sternocostal head of pectoralis major and complete absence of pectoralis minor muscles. Varying degrees of congenital malformations including, the other chest wall and upper limb muscles, breast, nipple and areola, rib cage, hand, and phalanges. Also skeletal, genitourinary, cardiac, and endocrine abnormalities have been associated. In the pathogenesis of the Poland's syndrome, the defective development of the blood supply of the subclavian, internal thoracic, and brachial arteries are accused. Poland's syndrome may be associated with Möbius and Klippel-Feil syndromes. These two syndromes are due to abnormalities of the primitive trigeminal arteries and the vertebral arteries, respectively.^[1]

This is the first case of Poland's syndrome associated with renal cell carcinoma. There is an association between Poland's syndrome and malignancy. Lymphoreticular malignancies,^[2] and solid tumors as leiomyosarcoma,^[3] neuroblastoma,^[4] Wilm's tumor,^[5] breast cancer,^[6] lung cancer,^[7] and gastric cancer^[8] in the cases with Poland's syndrome were reported. We, as the other authors assume that Poland's syndrome is a congenital disorder associated with increased risk of lymphoreticular and solid malignancies. The association between some congenital syndromes and malignancies is known. Down's syndrome and Bloom's syndrome have increased risks of leukemia. Also the associated syndromes with Poland's syndrome, as mentioned above, also have increased risks for the development of some tumors. Klippel-Feil syndrome has a risk for the development of the posterior fossa dermoid cysts^[9] and Möbius syndrome has a risk for the development of the Wilms' tumor.^[10]

MDCT is an important examination method for the trauma patients in emergency departments as our patient. The unconscious trauma patients underwent MDCT, so that multiple systems are evaluated. However, MDCT is commonly using in radiology practice for the detection of the chest. When the clinicians are suspected about a lung disease, for example unilateral hyperlucent lung at radiography, as our patient, the second imaging method is MDCT. Thus, the radiologists are commonly encountering with developmental abnormalities of the chest. Especially for radiologist, it is important to know the association between this syndrome and various tumors.

Therefore, all the examinations of the cases with Poland's syndrome should be carefully detected for a possible malignancy by the radiologist, and the incidental cases with Poland's syndrome should be aware of oncologic attention by the radiologists and clinicians.

REFERENCES

1. Fokin AA, Robicsek F. Poland's syndrome revisited. *Ann Thorac Surg* 2002;74:2218-25.
2. Sackey K, Odone V, George SL, Murphy SB. Poland's syndrome associated with childhood non-Hodgkin's lymphoma. *Am J Dis Child* 1984;138:600-1.
3. Shaham D, Ramu N, Bar-Ziv J. Leiomyosarcoma in Poland's syndrome. A case report. *Acta Radiol* 1992;33:444-6.
4. Caksen H, Patiroglu T, Ozdemir MA, Patiroglu TE, Poyrazoglu MH, Tercan M. Neuroblastoma and Poland syndrome in a 15-year-old boy. *Acta Paediatr Jpn* 1997;39:701-4.
5. Athale UH, Warrier R. Poland's syndrome and wilms tumor: An unusual association. *Med Pediatr Oncol* 1998;30:67-8.
6. Fukushima T, Otake T, Yashima R, Nihei M, Takeuchi S, Kimijima II, et al. Breast cancer in two patients with Poland's syndrome. *Breast Cancer* 1999;6:127-30.
7. Ahn MI, Park SH, Park YH. Poland's syndrome with lung cancer. A case report. *Acta Radiol* 2000;41:432-4.

8. Kurt Y, Demirbas S, Uluutku AH, Akin ML, Celenk T. Poland's syndrome and gastric cancer: Report of a case. *Eur J Cancer Prev* 2006;15:480-2.
9. Kennedy PT, McAuley DJ. Association of posterior fossa dermoid cyst and Klippel-Feil syndrome. *AJNR Am J Neuroradiol* 1998;19:195;discussion 196.
10. Yaris N, Aynaci FM, Kalyoncu M, Odemis E, Okten A. Wilms' tumor in a case with Mobius' syndrome associated with arthrogyposis and mega cisterna magna. *J Child Neurol* 2004;19:67-70.

Cite this article as: Yuksekkaya R, Celikyay F, Deniz C, Acu B. Poland syndrome and renal cell cancer. *Clin Cancer Investig J* 2012;1:77-9.
Source of Support: Nil, **Conflict of Interest:** None declared.

New features on the journal's website

Optimized content for mobile and hand-held devices

HTML pages have been optimized of mobile and other hand-held devices (such as iPad, Kindle, iPod) for faster browsing speed.

Click on [**Mobile Full text**] from Table of Contents page.

This is simple HTML version for faster download on mobiles (if viewed on desktop, it will be automatically redirected to full HTML version)

E-Pub for hand-held devices

EPUB is an open e-book standard recommended by The International Digital Publishing Forum which is designed for reflowable content i.e. the text display can be optimized for a particular display device.


Click on [**EPub**] from Table of Contents page.

There are various e-Pub readers such as for Windows: Digital Editions, OS X: Calibre/Bookworm, iPhone/iPod Touch/iPad: Stanza, and Linux: Calibre/Bookworm.

E-Book for desktop

One can also see the entire issue as printed here in a 'flip book' version on desktops.

Links are available from Current Issue as well as Archives pages.

Click on  View as eBook