Primary Hydatid Cyst of the adrenal gland: A case report and a review of the literature

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ABSTRACT

INTRODUCTION: In North Africa which is an endemic region for Hydatid Cyst, Tunisia is considered as an endemic country. The liver and lungs are common locations for Hydatid Cysts, whereas the Adrenal Glands are unusual and rare locations.

PRESENTATION OF CASE: Here is a report of primary Hydatid Cyst in a 55-year-old patient, with left hypochondrium pain as chief complaint. No remarkable findings were revealed by physical examination and blood analysis showed normal range. Hydatid serology was negative. The diagnosis of Hydatid Cyst was suspected based on CT Scan results which showed a well-circumscribed, non-enhanced, multicystic, 12 cm mass with scattered calcifications located in the left adrenal gland. Therefore, the patient underwent an open surgery with resection of the protruding dome of the cyst as it was attached to the renal pedicle, the pancreatic tail, the spleen and the jejunum. The final pathological examination of the specimen led to a Hydatid Cyst.

DISCUSSION: Throughout an extensive literature review that we have made, we have analyzed 54 reported cases, with their clinical presentations, biological exams, radiological features and surgical managements. The treatment should be surgical and has to be as conservative as possible. The prevention of the parasite transmission has to be the cornerstone of the disease management.

CONCLUSION: The hydatid cyst of the adrenal gland remains a rare diagnosis that has to be evoked in case of an adrenal gland cyst, especially in an endemic country.

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1. Introduction

The hydatid cyst is a well-known zoonosis that is endemic in some areas of the world, such as North Africa and Middle East, Central Asia, Australia and Latin America [1]. It is caused by Echinococcus granulosus larva, after being infested with the parasite through ingestion of contaminated water or vegetables, or direct contact with dogs. Common locations of the hydatid cyst are the liver and the lungs. Adrenal gland is an unusual and extreme rare location of the hydatid cyst, especially when it is the primary location. Herein we report a case of a primary hydatid cyst of the adrenal gland. Clinical radiological features will be discussed as well as the surgical procedure with a literature review of the previous cases. The work has been reported in line with the SCARE criteria [2].

2. Case report

A 55-year-old patient, living in an endemic area, was referred to our institution for pain in the left hypochondrium evolving for 6 months, with no other symptoms such as: nausea and vomiting. Arterial blood pressure was within normal limits. No past medical history was reported. No anomalies were detected in the physical examination. Blood analysis including complete blood count, 24-h urinary VMA and metanephrine were normal. Serological analysis of the blood was negative for Echinococcus IgM and IgG. An abdominal sonography showed a large hypoechoic retroperitoneal mass in the upper pole of the left kidney containing internal cystic component. The CT scan showed a well-circumscribed non-enhanced multi-cystic 12 cm mass with scattered calcifications located above the left kidney and the left adrenal gland which seemed shadowed (Fig. 1). The mass has a compressive effect on the adjacent organs.
with no perirenal or adrenal fat infiltration, no invasion of thick wall blood vessels and no extension beyond renal lodge, suggesting the diagnosis of hydatid cyst. There were no other locations, particularly in the liver or the lungs. Subsequently, the patient underwent a surgical removal of the mass by an open laparotomy by a left subcostal incision. Intraoperatively, we found a cyst measuring 12 cm in its largest diameter, contiguous to the renal pedicle, the pancreatic tail, the spleen, and the jejunum. Its wall colour was pearly white. We could surgically release the mass from the kidney and the jejunum, but the dissection along the pancreas was dangerous. Thus, we performed a resection of the protruding dome, after sterilizing the content of the cyst (Fig. 2). The puncture of the cyst showed a multivesicular clear content. The postoperative course was uneventful and the patient was discharged on the 3rd day post-
### Table 1

Reported cases of hydatid cyst of the adrenal gland.

| Author               | Age | Sex | Environment | Contact with animals | Symptoms                        | Hypereosinophilia | Serology | Side | Size on imaging (cm) | Primary/Secondary | Treatment by albendazole | Surgical access | Follow up (months) | Recurrence |
|----------------------|-----|-----|-------------|----------------------|---------------------------------|--------------------|----------|------|---------------------|------------------|----------------------|----------------|-------------------|------------|
| Fitzgerald EJ (1985) | 48  | M   | N/A         | N/A                  | Right hypochondruim pain       | N/A                | Positive | Right | 18                  | Primary          | N/A                  | Open surgery    | N/A               | N/A        |
| Strojinsky AS (1990) | 50  | N/A | N/A         | N/A                  | N/A                             | N/A                | N/A      | N/A  | N/A                 | Secondary        | N/A                  | Open surgery    | N/A               | N/A        |
| Schroeter G (1998)   | 48  | M   | Rural       | Yes                  | Right hypochondruim pain       | No                 | Negative | Right | 9.5                 | Primary          | No                   | Open surgery    | N/A               | N/A        |
| Defechereux T (2000) | 37  | F   | N/A         | N/A                  | Left Flank Pain                 | N/A                | N/A      | Left  | 5                   | Secondary        | No                   | Laparoscopy     | N/A               | N/A        |
| C.G. Yenyl (2000)    | 51  | F   | N/A         | N/A                  | Left Flank pain                 | N/A                | Positive | Left  | 6                   | Primary          | N/A                  | N/A            | N/A               | N/A        |
| et el-Irissi Dalahi A (2002) | 28 | N/A | N/A         | No                   | Right hypochondruim pain       | Yes                | Positive | Left  | 7                   | Primary          | No                   | Open surgery    | N/A               | N/A        |
| Escudero MD (2002)   | 40  | F   | Urban       | No                   | Hypertension                    | N/A                | N/A      | Left  | 10                  | Primary          | N/A                  | Open surgery    | 12                | No         |
| 53  | F   | N/A | N/A         | N/A                  | Right hypochondruim pain       | N/A                | Positive | Right | 8                   | Primary          | N/A                  | N/A            | N/A               | 16         |
| Mulfide (2003)       | 18  | N/A | Rural       | No                   | Hypochondruim                  | N/A                | Positive | Right | 12                  | Primary          | N/A                  | N/A            | N/A               | 16         |
| Nuran (2004)         | 28  | F   | N/A         | No                   | Right hypochondruim pain       | N/A                | Positive | Right | 5                   | Secondary        | Yes                  | N/A            | 16                | No         |
| Alkay (2004)         | 48  | F   | N/A         | N/A                  | Right Flank pain                | N/A                | Positive | Right | 20                  | Primary          | N/A                  | N/A            | 16                | No         |
| Gürdal M (2004)      | 47  | F   | N/A         | N/A                  | Left Flank pain                 | N/A                | Negative | Right | 6                   | Primary          | N/A                  | Open Surgery    | 12                | No         |
| Recai Gurbuz (2005)  | 20  | M   | N/A         | No                   | Right hypochondruim pain       | N/A                | Positive | Right | 5.6                 | Primary          | N/A                  | Open Surgery    | 24                | No         |
| Bedioui (2005)       | 50  | M   | N/A         | No                   | Left hypochondruim pain         | No                 | Negative | Left  | 6                   | Primary          | N/A                  | Open surgery    | 36                | No         |
| Ozarmagan S (2006)   | 54  | F   | N/A         | No                   | Hypertension                    | N/A                | Positive | Left  | 5                   | Primary          | Yes                  | Open Surgery    | N/A               | No         |
| Saffi-Fath Elzehba (2006) | 61 | F   | Yes         | Yes                  | Hypertension                    | Yes                | Positive | Left  | 5.8                 | Primary          | Yes                  | Open Surgery    | N/A               | No         |
| Tsaroucha AK (2007)  | 56  | M   | Rural       | Yes                  | Hypertension                    | No                 | Negative | Left  | 7                   | Primary          | No                   | Open surgery    | 12                | No         |
| Shintaro Maru (2007) | 79  | F   | Urban       | Yes                  | Impaired General Condition      | No                 | Negative | Left  | 5.5                 | Primary          | No                   | Open Surgery    | N/A               | No         |
| Gianilorenzo Dionigi (2007) | 68 | F   | N/A         | Yes                  | Left Flank pain                 | No                 | Positive | Left  | 3                   | Primary          | Yes                  | Laparoscopy     | 6                 | N/A        |
| Ruiz-Rabelo JF (2008) | 70  | F | N/A         | Yes                  | Left hypochondruim pain + Fever | No                 | Negative | Left  | 9                   | Secondary        | No                   | Open surgery    | No                 | N/A        |
| Tamilsas Kamishuma (2009) | 77 | F   | N/A         | Yes                  | Hypertension                    | N/A                | Right    | 6                   | Primary          | N/A                  | Laparoscopic     | 13                | No         |
| O.Barakiet (2010)    | 38  | F   | Rural       | Yes                  | Left Flank pain                 | N/A                | N/A      | Left  | 7                   | Primary          | N/A                  | Open surgery    | 36                | No         |
| B Geramizadeh (2011) | 49  | F   | N/A         | Yes                  | Left Flank pain + Hypertension  | No                 | N/A      | Left  | 8.2                  | Primary          | No                   | N/A            | 2                 | No         |
| Limaeni F (2012)     | 55  | F   | N/A         | No                   | Left hypochondruim pain + Hypertension | N/A     | N/A      | Left  | 12                  | Secondary        | N/A                  | N/A            | No                 | No         |
| Fadli Tazi (2012)    | 64  | M   | N/A         | Yes                  | Right hypochondruim pain + Hypertension | Negative  | Yes      | Left  | 14.5                 | Primary          | Yes                  | Open Surgery    | No                 | No         |
| Maral Mokhtari (2012) | 66 | F   | N/A         | Yes                  | Right Flank pain + Hypertension | N/A                | N/A      | Right | 5                   | Primary          | No                   | Open Surgery    | N/A               | No         |
| Huang (2013)         | 45  | M   | N/A         | No                   | Hypertension                    | N/A                | Right    | 9.5                  | Primary          | No                   | Open surgery    | 24                | N/A        |
| aëkkalla Darwish (2013) | 30 | F   | N/A         | No                   | Hyperemesis gravidarum          | No                 | Negative | Right | 12                  | Primary          | No                   | Open surgery    | N/A               | No         |
| Babinski A (2014)    | 47  | F   | N/A         | No                   | Hypertension                    | Yes                | N/A      | Right | 6.8                  | Primary          | No                   | Laparoscopic     | 12                | No         |
| Santosh Kumar (2014) | 51  | F   | N/A         | Yes                  | Left hypochondruim pain         | No                 | Positive | Left  | N/A                  | Primary          | Yes                  | Open Surgery    | 6                 | No         |
| Afsin Mohammadi (2014) | N/A | M   | N/A         | Yes                  | Hypertension                    | N/A                | N/A      | Left  | 13                  | Primary          | Yes                  | Open Surgery    | 6                 | No         |
| Walter Nardi (2015)   | 55  | M   | N/A         | Yes                  | Left hypochondruim pain         | No                 | Positive | Left  | 6.5                  | Primary          | No                   | Laparoscopic     | N/A               | No         |
| Ammar Mahmoudi (2015) | 76  | F   | Rural       | No                   | Right hypochondruim pain        | N/A                | N/A      | Right | 5                   | Secondary        | No                   | Open surgery    | 24                | No         |
| Silke Spath (2016)    | 78  | M   | N/A         | No                   | Right hypochondruim pain        | No                 | Positive | Left  | 7.4                  | Primary          | Yes                  | Open surgery    | 5                 | No         |
| Gaurav Prakash (2016) | 35  | M   | Rural       | Yes                  | Right Flank pain                | N/A                | Negative | Right | 16                  | Primary          | Yes                  | Open surgery    | N/A               | No         |
| Fatehi Elnour Elzein (2016) | 44 | M   | N/A         | Yes                  | Right Flank pain                | N/A                | Negative | Right | 10                  | Primary          | Yes                  | Open Surgery    | N/A               | No         |
| Sami Akboulut (2016)  | 64  | M   | N/A         | No                   | Vague abdominal pain            | No                 | Positive | Left  | 15                  | Primary          | Yes                  | Open Surgery    | 24                | No         |
| Giovanni Aprea (2016) | 78  | F   | N/A         | No                   | Right Flank pain                | N/A                | Positive | Left  | 3.4                  | Primary          | Yes                  | Laparoscopy     | 24                | No         |
| Ann-Katrin Seidel (2017) | 16 | M   | N/A         | No                   | Hypochondruim                  | N/A                | Positive | Right | 7                   | Primary          | Yes                  | Open surgery    | 24                | No         |
| Sami Akboulut (2018)  | 16  | M   | Urban       | No                   | Right hypochondruim pain        | N/A                | Positive | Right | 7                   | Primary          | Yes                  | Open surgery    | 12                | No         |
| Present case (2020)   | 55  | M   | Urban       | No                   | Right hypochondruim pain        | N/A                | Negative | Right | 12                  | Primary          | No                   | Open surgery    | 12                | No         |

N/A: not available.
3. Discussion

Hydatid cysts are usually found in the liver or the lungs. It occurs when humans ingest accidentally echinococcus eggs found in the dogs’ stools. After passing through the digestive system, the larvæ are released; they pass through hepatic filter to the lungs. If they are not destroyed by the immune system or trapped in the filtration system; they can spread anywhere in the body, giving unusual locations of the hydatid cyst as spleen, kidneys, brain, bones, heart, muscle tissue, pancreas, retroperitoneum, thyroid and adrenal glands, through the arterial circulation. Other mechanisms of the dissemination of the larvae imply entering the systemic circulation through lymphatic vessels, or by adjacent contact.

The adrenal location of the hydatid cyst is rare and accounts for less than 0.5% off all hydatid cysts [3,4]. It generally occurs in the context of a disseminated hydatid disease. In the other hand, hydatid cyst constitutes 7% of adrenal gland cysts. Isolated, primary adrenal hydatid cyst are extremely rare [5,6]. Most of the published cases are case reports. The largest series published in the literature was made by Ackay [7] in 2004 with 9 reported cases. In 2006, Horchani et al. have reported 6 cases over a 10 year period [8].

Based on a systematic Pubmed search using the keywords ‘Adrenal Gland’ and ‘Hydatid Cyst’, we have found 76 cases published in the literature. We have excluded those who could not be opened. We have selected therefore 54 cases for review. All the cases are summarized in [Table 1]. The data has been analyzed using SPSS version 19.0 software. The main limitation of our analysis is the lack of information in some cases.

Adrenal hydatid is more likely to be discovered during the 5th decade. The mean age in our review was 50 years. It occurred equally in men and women. It is most frequently discovered incidentally during surgery or on radiology, as autopsies reports shows an incidence between 0.06 and 0.18% with 92% of the lesions being unilateral [9]. Common presenting symptoms may include vague hypochondrium or flank pain, nausea and vomiting, or palpable mass. Rarely, is discovered by arterial hypertension, resulting from compression of renal parenchyma in large cysts [10]. In our review, abdominal or hypochondrium pain was the most presenting symptom in 66% of the cases. Arterial hypertension occurred in 17% while it was discovered incidentally in 11% of the cases. The right side was most affected in 62% of the cases. Biological exams could help raise the diagnosis, but they are unspecific. Hypereosinophilia was reported 4 times while hydatid serology was positive in 20 cases. Of those cases, the hydatid cyst was secondary to a previous location in 3 patients. In our patients, we did not find hypereosinophilia, and the hydatid serology was negative.

The radiological findings can orient the diagnosis. Ultrasound remains the first exam performed for this localization. The depth of the adrenal glands and sometimes peripheral calcifications make this exploration difficult. The computed tomography scan then allows for a better understanding of the location and relationships with the surrounding organs [11]. But often, the diagnosis assessment is made intraoperatively, as several other diagnoses can be evoked. Differential diagnosis may include cystic lymphangioma, pseudo haemorrhagic cyst, cyst with epithelial coating or extra-adrenal cystic masses. The standard treatment of the hydatid cyst remains surgical.

The most recommended surgical procedure is the resection of the protruding dome with drainage of residual cavity. A pericystectomy of the hydatid cyst, or, if this is not possible, total excision of the adrenal gland can also be performed. Either laparoscopic or open procedure can be used [12,13], depending on the tumor size. Laparoscopic procedure is usually avoided, especially when the tumor size is above 6 cm and when radiological findings show daughter cysts within the cavity because of the risk of peritoneum spread due to insufflation [13]. In our review, laparoscopic method was used in 9 cases, and open procedure in 33 cases.

Laparotomy can be performed either by intercostal lumbar access with or without resection of the rib; or transperitoneal anterior access allowing a sufficient view on the liver in case of associated localization. In our case, we performed a left subcostal incision, finding an accolated cyst to the pancreas, jejunum and renal pedicle. It is important to protect the operative field with compresses soaked in hypertonic (20%) sodium chloride solution to prevent larvae dissemination in case of accidental opening of the cyst intraoperatively. Conservative management of the adrenal gland is the standard attitude, except in the case of adrenal gland destruction by the cyst. In most of the cases, post-operative course is uneventful. Surgical removal of the hydatid cyst may normalise the hypertension if it was associated to the hydatid cyst preoperatively. We found normalization of the hypertension in 63% in our review. Among the published cases, no recurrence has been reported, as the case of our patient 10 months after the surgery. Post-operative albendazole prophylaxis is still controversial, even if some authors recommend it. The prevention of hydatid transmission remains an indispensable measure to avoid hydatid disease whatever his location is.

4. Conclusion

The hydatid cyst of the adrenal gland remains a rare diagnosis that has to be evoked in case of an adrenal gland cyst, especially in an endemic country. Clinical presentation and physical examination remain unspecific. CT Scan in combination with hydatid serology help raise the diagnosis, that is confirmed by the final pathological examination of the specimen. The treatment is surgical and has to be as conservative as possible. The prevention of the parasite transmission has to be the cornerstone of the disease management.

Declaration of Competing Interest

The authors have no conflict of interest to declare.

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Ethical approval

Given the nature of the article, a case report, no ethical approval was required.
Consent

Written informed consent was obtained from the patient for publication of this case and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

- Skander Zouari: Writing - original draft
- Marouene Chakroun: Writing - review & editing.
- Hana Bibani: Writing - original draft.
- Ahmed Saadi: Writing - review & editing.
- Linda Haj Kacem: Data interpretation of the radiological findings.
- Ahlem Bظل: Data interpretation of the pathological findings.
- Soumaya Rammeh: Data interpretation of the pathological findings.
- Haroun Ayed: Supervision; Reviewing and editing
- Mohamed Chebil: Supervision; Reviewing and editing

Registration of research studies

This does not apply as it is a case report of a patient who has given written consent and has been de-identified. It is therefore not prospective research involving human participant.

Guarantor

Dr. Skander Zouari.

Provenance and peer review

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