ISSN: 2637-4617

Research Article

Current Opinion in Gynecology and Obstetrics

Primary Ovarian Choriocarcinoma: One Case in 20 Years at INCA and Literature Review

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Received date: November 02, 2018; Accepted date: January 21, 2019; Published date: January 26, 2019

Abstract

Primary ovarian choriocarcinoma treatment doesn't have a specific protocol and therefore several questions about this disease are still unanswered. This study is interesting because we performed a comprehensive review of the medical literature and report through descriptive analysis the only case registered as Primary ovarian choriocarcinoma in the period of 20 years at the Brazilian National do Cancer. Reading this paper may offer you the opportunity to understand biological behavior, pathological and clinical aspects, imaging indications and usefulness for a rare and deadly disease. Because it is a rare neoplasm and with few studies, there is no consensus for the treatment.

Keywords: Choriocarcinoma, Ovarian neoplasms, Medical oncology

Introduction

Primary ovarian choriocarcinoma is an extremely rare neoplasm and therefore little reported in the medical literature. It comprises 2.1% of all ovarian germ cell neoplasms and accounts for only 0.6% of all neoplasms of ovarian origin [1-5]. It can have either gestational or non-gestational origin. The gestational type is defined as the result of the proliferation of an uterine or tubal trophoblastic disease to the ovary or, more rarely, the anomalous development of a gravid ovary. However, the non-gestational form results from the anomalous development of the ovary's own germinative cells without gestation [6-10]. However, the distinction between Ovarian choriocarcinoma presentation forms is extremely difficult due to immunohistochemical similarity [8]. Some authors establish tumor DNA analysis as the ideal method for this distinction [2-7], however, this technique is little used due to its high cost [2-9].

Clinical manifestation of the primary ovarian choriocarcinoma is varied ranging from pelvic pain, bleeding during intercourse, dyspareunia and even acute abdomen or cor pulmonale due to tumor embolism [5]. Due to this variable manifestation and to the unfavorable prognosis, primary ovarian choriocarcinoma is characterized as an oncological urgency that requires immediate diagnosis and therapeutic approach [1-5,11].

The rarity of Primary ovarian choriocarcinoma was the determining factor to the accomplishment of this study that aims to review the medical literature on epidemiological, pathological and clinical aspects of this lesion.

Clinical presentation

Between January 1991 and March 2016, the Brazilian National do Câncer (INCA), located in the city of Rio de Janeiro, Brazil, admitted approximately 3950 patients diagnosed with ovarian tumor, and among all these cases, only one case of primary ovarian choriocarcinoma was found, after approval by the Ethics and National Research Committee.

This single case concerns a 35-year-old white female patient admitted to Onco-gynecology Division of this hospital due to a pelvic mass in January 2016.

Lab and radiological investigations

The patient brought, with herself, the result of a Magnetic Resonance Imaging (MRI) of the pelvis, carried out at our institute dated December 2015. This MRI showed heterogeneous expansive formation with necrotic area located in the right adnexal region measuring 7.5 × 7.8 × 6.4 cm and estimated volume of 187.2 cm^3. In addition, serum levels of βHCG were 831,659.0 mIU/mL, CA125 was 42.22 U/mL and CEA was 7.21 ng/mL at that time. At the initial examination, the patient presented Performance Status (PS) 2, (Table 1) tachydyspnea, abdomen with a fixed, hard and palpable mass with irregular contours occupying the area from mesogastrium to hypogastrium and projected to both iliac fossae, especially to the right one.

The patient was referred to hospital admission due to respiratory distress associated with elevated βhCG and underwent diagnostic investigation. Computed tomography (CT) scan of the chest, abdomen and pelvis, performed at our institute, revealed the presence of multiple pulmonary implants, as well as massive heterogeneous pelvic expansive formation with cystic and solid components measuring 21.2 × 13 × 16.5 cm, extending to umbilical region and pushing intestinal loops laterally, densification of mesenteric fat and absence of free liquid (Figures 1, 2 and 3).

Abdomen/pelvis CT scan showing heterogeneous expansive formation on topography of right attachment on 02/01/2016 (figure 1, 2).

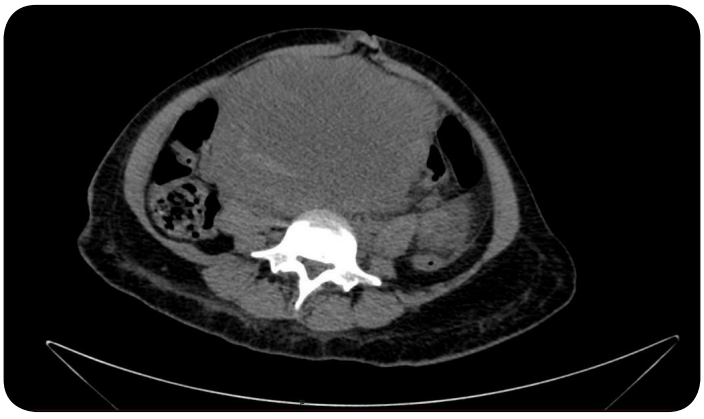

Figure 1: Abdomen/pelvis CT scan.

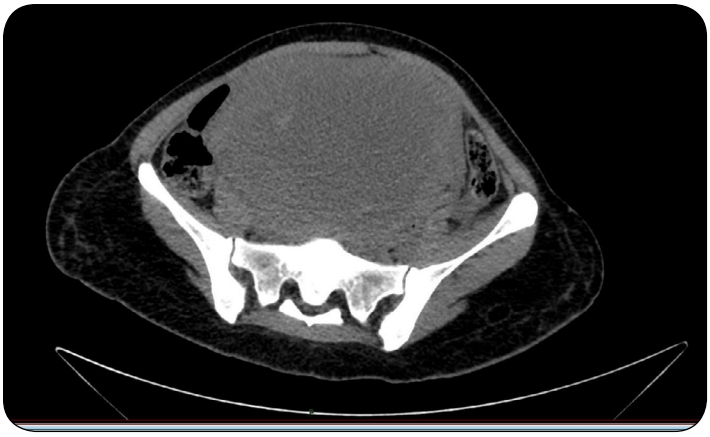

Figure 2: Supplementary material

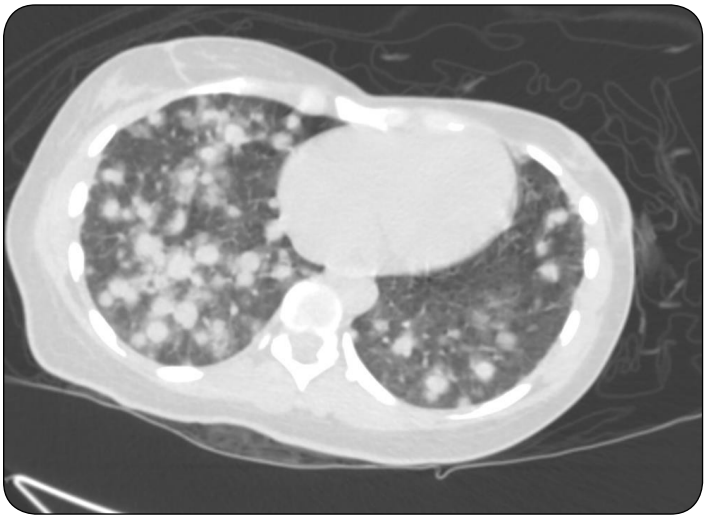

Figure 3: Thorax CT scan evidencing multiple secondary implants on 02/01/2016.

Diagnosis

On the second day of the hospitalization, the patient evolved with a significant clinical respiratory worsening. Therefore, the surgical procedure for diagnostic confirmation was suspended and a percutaneous biopsy guided by pelvic CT scan was performed and add to this, an urgent histopathological evaluation was requested due to the strong suspicion of ovarian choriocarcinoma.

Treatment and complications

Still on the second day of hospitalization, chemotherapy was started with cisplatin and etoposide. On the third day of hospitalization, the patient evolved with Acute Respiratory Insufficiency (ARI) requiring oro-tracheal intubation and Mechanical Ventilation (MV) in the Intensive Care Unit (ICU). On the same day, percutaneous biopsy histopathological report was compatible with choriocarcinoma with positive immunohistochemistry for CK7 and βHCG, consequently the patient underwent EMA/

Table 1: Supplementary material - Performance status (PS).

Zubrod Scale (ECOG)	Karnofsky scale (%)
PS 0 - Normal activity	100 - no complaint: no evidence of the disease
	90 - able to have a normal life; minor signs or symptoms of the disease
PS 1 - Symptoms of the disease, but can walk and have a normal daily life	80 - some signs or symptoms of the disease upon effort
	70 - able to take care of him/herself; unable to carry out normal activities or perform active work
PS 2 - Out of bed more than 50% of the time	60 - needs occasional assistance, but is still able to accomplish most of his/her activities
	50 - requires substantial assistance and frequent medical care
PS 3 - In bed more than 50% of the time, in need of more intensive care	40 - incapable; requires special care and assistance
	30 - very incapable; hospitalization recommended although death is not imminent.
PS 4 - Confined to bed	20 - very weak; requires hospitalization and active supportive care
	10 - moribund, lethal processes progressing rapidly

Source: CONDUTAS DO INCA/MS / INCA/MS PROCEDURES - Cuidados Paliativos Oncológicos - Controle de Sintomas - Revista Brasileira de Cancerologia, 2002, 48(2): 191–211

CO chemotherapy (etoposide, methotrexate, actinomycin D at stage 1 and cyclophosphamide and vincristine at stage 2). The patient remained on MV for 11 days and was discharged from the Intensive Care Unit 7 days after oro-tracheal extubation. βHCG serum measured after the second cycle of chemotherapy showed a significant decrease in relation to the value presented at the time of admission (βHCG after chemotherapy: 230,995 mUI/mL). In abdominal CT scan performed in March 2016, still during hospitalization referred, a massive subcapsular expansive formation with heterogeneous appearance was observed at the right hepatic lobe, associated with the appearance of other small nodules scattered over the hepatic parenchyma. In MRI of the skull performed in March 2016, still during hospitalization referred, staging examination, was observed the presence of multiple nodules lesions, distributed diffusely over the cerebral and cerebellar parenchyma, two of which were located in the occipital lobes, the largest on the left, measuring 1.1 cm, with signs suggestive of late subacute bleeding. The patient was discharged after 48 days of hospitalization with PS 1.

Response to treatment or progress of disease

EMA/CO protocol was followed for 4 cycles up to April 2016, and because of the hepatotoxicity, evidenced by laboratory tests in April 2016, a second line of TE/TP chemotherapy regimen (paclitaxel-etoposide and paclitaxel-cisplatin) was started and maintained for 4 cycles up to August 2016, when, during the outpatient follow-up, the tests showed elevated serum levels of βHCG and raised the hypothesis of disease progression to the Central Nervous System. Skull CT scan of September 2016 revealed the appearance of two hyperdense, nodular lesions with dural basis of implantation in the right occipital lobe, the smaller one measuring 0.4 cm and the larger one measuring 0.9 cm, the latter with adjacent vasogenic edema compatible with secondary implants. Although throughout this period the patient was PS 2, in the presence of uncontrolled hepatic and pulmonary metastatic disease and in the absence of neurological symptoms, the patient was not referred to radiotherapy evaluation of the cerebral lesion, and due to the elevation of βHCG and CT scan findings, the third line of chemotherapy with FA was started in August 2016 (5-fluoracil and actinomycin D).

Outcomes

However, on October 17, 2016, the patient was admitted to the emergency room with a left unilateral headache associated with left inferior quadrantanopsia in the left eye. Skull CT scan revealed increased expansive lesions in the Central Nervous System with a midline deviation of 0.6 cm to the left (Figures 4 and 5) evolving to death on October 18, 2016.

Skull CT scan on 10/17/16 evidencing progression of the disease to the Central Nervous System (figure 4, 5).

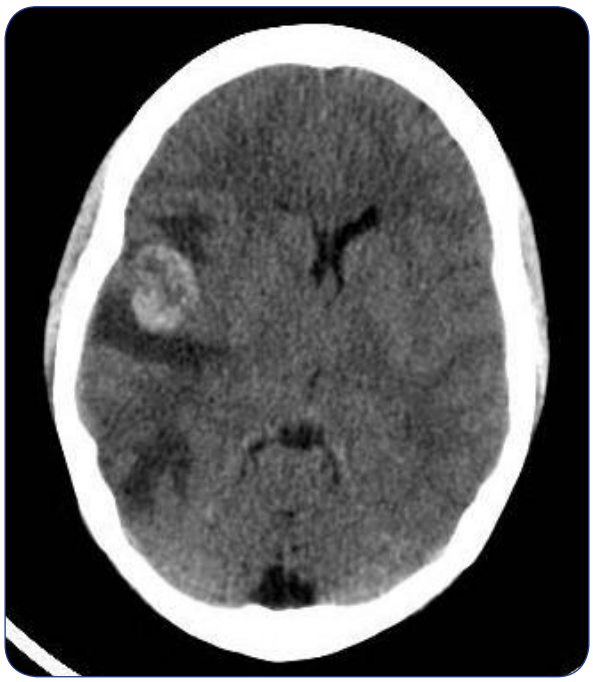

Figure 4: Skull CT scan.

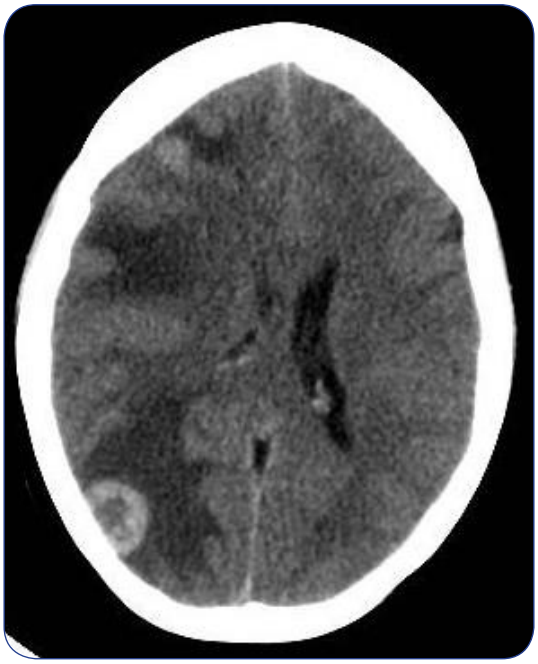

Figure 5: Supplementary material.

Discussion and Conclusion

Epidemiology of the cancer

Primary ovarian choriocarcinoma is an extremely rare neoplasm and occurs more frequently in women with childbearing potential [1-8]. In women with active sex life, due to the high level of βHCG, the differential diagnosis between ectopic pregnancy and abortion should be considered. Due to the rarity of reported cases of ovarian choriocarcinoma, the ethnic prevalence for the occurrence of this pathology cannot yet be stated [1-6].

Cell of origin, pathological features and tumor biology

The real mechanism for the development of this neoplasm is not clear, but it is known that it can originates from gestational tissue (gestational type) or ovarian origin germ cells (non-gestational type) [1]. Many researchers state that in virgin women diagnosed with choriocarcinoma it is possible to declare the non-gestational form without confirmation by DNA analysis [8].

Most common clinical resentation

However, although the gestational form is more frequent (1 case for 369,000,000 pregnancies), the non-gestational form also occurs in women with childbearing potential, and even after menopause [12,13]. Usually, previous history of molar gestation, tubal gestation and abortion is associated with gestational presentation of this type of neoplasm. However, some studies argue that the distinction between the two manifestations is surely proven through tumor DNA analysis, but the high cost of this method limits its routine application [8].

Recommended/proposed treatment options

Because it is a rare condition with only 65 cases of primary ovarian choriocarcinoma reported in the medical literature (Tables 2 and 3), many questions about the staging and treatment of Primary ovarian choriocarcinoma remain unanswered [5]. Aucouturier et al. [14], defend surgical staging and recommend total hysterectomy with bilateral salpingo-oophorectomy, omentectomy, peritoneal biopsies, and lymphadenectomy. Santos et al. [5], recommend total hysterectomy with bilateral salpingo-oophorectomy. Heo et al. [4], also recommend surgical staging, and defend performing total abdominal hysterectomy with bilateral salpingo-oophorectomy, omentectomy, peritoneal biopsies of diaphragmatic domes and parieto-colic gutters with peritoneal lavage collection, and suggest that lymphadenectomy should be performed only when lymph node metastasis is suspected. Exman et al. [1], Xin et al. [8] and Wang et al. [12], recommend imaging staging, and the treatment for cases with distant metastasis should begin with neoadjuvant chemotherapy followed by total hysterectomy with bilateral salpingo-oophorectomy, omentectomy, peritoneal biopsies and lymphadenectomy. In young patients with tumor limited

Table 2: Non-gestational primary ovarian choriocarcinoma-literature review.

					Surgery			
Author / Reference / Year	**Age**	**Initial Beta HCG**	**Diagnostic**	**Stage**	**Via**	**Surgical Description**	**Chemotherapy regimen**	**Prognosis**
Exman [1] 2006	24	675,713	Non-Gestational	Unknown	Unknown	TAH; BSO	BEP	1 month without disease evidence
Hirabayashi [2] 2006	50	704.1	Non-Gestational	Unknown	Laparotomy	TAH + BSO + pelvic lymphadenectomy	unknown	death
Xin [5] 2015	23	unknown	Non-Gestational	IIB	Laparoscopic	LSO + omentectomy + pelvic and paraortic lymphadenectomy + peritoneal biopsies	BEP	9 months without disease evidence
Tsujioka [7] 2003	19	110,000	Non-Gestational	IV	Laparotomy	LSO + partial omentectomy + right ovarian biopsy	EMA-CO	unknown
Choi [9] 2013	33	74,612	Non-Gestational	Unknown	Laparoscopic	LSO, multiple biopsies	EMA	5 years without disease evidence
Hayashi [10] 2015	10	6,600	Non-Gestational	Unknown	Laparotomy	RSO	BEP	5 years without disease evidence
Wang [12] 2016	13	unknown	Non-Gestational	Unknown	Laparotomy	TAH + BSO	PVB	death
Rao [14]2015	26	8,160	Non-Gestational	Unknown	Laparotomy	RSO + omentectomy + splenectomy + partial adrenalectomy	BEP	1 year without disease evidence
LV [15] 2011	48	7,664.30	Non-Gestational	Unknown	Laparotomy	BSO, TAH, pelvic lymphadenectomy, omentectomy, appen-dectomy	BEP	1 year without disease evidence
Gon [16] 2010	21	2,790.00	Non-Gestational	Unknown	Unknown	RSO	unknown	unknown
Park [17] 2009	55	64,838	Non-Gestational	Unknown	Laparotomy	TAH + BSO, multiple biopsies	BEP	20 months with-out disease evidence
Wang [18] 2009	23	26,516	Non-Gestational	Unknown	Laparoscopic	TAH + BSO, pelvic lymphadenectomy	BEP	2 months without disease evidence
Kong [19] 2009	10	unknown	Non-Gestational	IC	Laparotomy	LSO + partial omentec-tomy	BVP	2 months without disease evidence
Mood [20] 2009	32	5,500	Non-Gestational	Unknown	Laparotomy	TAH + BSO + lymph node debulking	BEP + EMA-CE	5 years without disease evidence
Chen [21] 2008	23	unknown	Non-Gestational	IA	Laparoscopic	unknown	unknown	3 years without disease evidence
Gerson [22] 2007	33	564,000	Non-Gestational	Unknown	Laparoscopic	RSO	EMA-CO	1 year without disease evidence
Koo [23] 2006	33	185,000	Non-Gestational	Unknown	Laparoscopic	TAH + BSO + omentectomy + pelvic lymphadenectomy	MAC	no evidence of disease – un-known time
Bazot [24] 2004	38	2,460.00	Non-Gestational	Unknown	Unknown	TAH + BSO	unknown	7 years without disease evidence
Balat [25] 2004	24	8,968	Non-Gestational	Unknown	Unknown	TAH + BSO + pelvic lymphadenectomy + omentectomy	BEP	death

					Surgery			
Author / Reference / Year	**Age**	**Initial Beta HCG**	**Diagnostic**	**Stage**	**Via**	**Surgical Description**	**Chemotherapy regimen**	**Prognosis**
Ozdemir [26] 2004	13	91,028	Non-Gestational	Unknown	Laparotomy	RSO	MAC	9 months without disease evidence
Simard [27] 1937	17	positive	Non-Gestational	IIB	Laparotomy	RSO	unknown	death
Backus and Griffin [28] 1941	13	unknown	Non-Gestational	IIB	Laparotomy	TAH + BSO	unknown	death
Oliver and Horne [29] 1948	11	positive	Non-Gestational	I	Laparotomy	RSO	unknown	death
Groeber [30] 1963	13	unknown	Non-Gestational	IA	Laparotomy	LSO	unknown	death
DeHaan [31] 1965	7	200	Non-Gestational	IA	Laparotomy	RSO	unknown	19 months without disease evidence
Hay and Stewart [32] 1969	13	positive	Non-Gestational	IC	Laparotomy	RSO	MTX	15 months without disease evidence
Panayotou et al. [33] 1971	12	positive	Non-Gestational	I	Laparotomy	TAH + BSO	MTX	death
Smith et al. [34] 1973	7	high	Non-Gestational	Unknown	Laparotomy	unknown	MAC	8 months without disease evidence
Shah et al. [35] 1974	14	unknown	Non-Gestational	II	Laparotomy	autopsy	-	death
Adelman et al. [36] 1975	1	unknown	Non-Gestational	Unknown	Laparotomy	unilateral annexectomy	MAC	no evidence of disease -unknown time
Gerbie et al. [37] 1975	16	unknown	Non-Gestational	IV	Laparotomy	LSO	MTX	11 years without disease evidence
Gerbie et al. [37] 1975	17	unknown	Non-Gestational	IA	Laparotomy	RSO	MTX ActD	6 years without disease evidence
Gerbie et al. [37] 1975	17	unknown	Non-Gestational	III	Laparotomy	LSO	MAC	1 year without disease evidence
Stevens et al. [38] 1979	19	160,000	Non-Gestational	IV	Laparotomy	TAH + BSO	MTX ActD	death
Piver and Lurain [39] 1979	unknown	high	Non-Gestational	IC	Laparotomy	unknown	MTX ActD	8 months without disease evidence
Creasman et al. [40] 1979	unknown	unknown	Non-Gestational	III	Laparotomy	BSO	MAC	4, 5 years without disease evidence
Vance and Greisinger [41] 1985	9	34	Non-Gestational	IC	Laparotomy	RSO	VBP	6 months without disease evidence
Axe et al. [42] 1985	6	unknown	Non-Gestational	IC	Laparotomy	RSO	none	10 years without disease evidence
Axe et al. [42] 1985	11	positive	Non-Gestational	I	Laparotomy	RSO	none	death
Raju et al. [43] 1985	16	unknown	Non-Gestational	IV	Laparotomy	autopsy	none	death

					Surgery			
Author / Reference / Year	**Age**	**Initial Beta HCG**	**Diagnostic**	**Stage**	**Via**	**Surgical Description**	**Chemotherapy regimen**	**Prognosis**
Sengupta and Everett [44] 1987	11	unknown	Non-Gestational	Unknown	Laparotomy	unilateral oophorectomy	unknown	unknown
Pippitt et al. [45] 1988	unknown	unknown	Non-Gestational	Unknown	Laparotomy	unilateral oophorectomy	VAB-VI	9 months without disease evidence
Spingler et al. [46] 1990	20	positive	Non-Gestational	Unknown	Laparotomy	unknown	unknown	death
Gribbon et al. [47] 1992	unknown	high	Non-Gestational	Unknown	Laparotomy	unknown	unknown	death
Gribbon et al. [47] 1992	11	high	Non-Gestational	IIC	Laparotomy	unknown	unknown	1 year without disease evidence
Brown et al. [48] 1993	11	unknown	Non-Gestational	Unknown	Laparotomy	unilateral annexectomy	unknown	2 years without disease evidence
Trigueros et al. [49] 1995	21	200,000	Non-Gestational	III	Laparotomy	TAH + BSO	VBP	4 years without disease evidence
Chou et al. [50] 1997	39	71,885	Non-Gestational	IV	Laparotomy	TAH + BSO	Carboplatin, etoposide phos-phamide	17 months with-out disease evidence
Goswami et al. [51] 2001	18	88,385	Non-Gestational	IA	Laparotomy	LSO + right cystectomy	MAC	5 months without disease evidence
Shin et al. [52], 1994	45	132,005	Non-Gestational	Unknown	Laparotomy	TAH + BSO	MAC	1 year without disease evidence
Byeun et al. [53], 1995	28	13,378	Non-Gestational	Unknown	Laparotomy	RSO	EMA-CO	1 year without disease evidence
Kim et al. [54], 1997	16	565,000	Non-Gestational	Unknown	Laparotomy	TAH + BSO	MAC	death
Chien et al. [55], 2004	21	1787.052.3	Non-Gestational	Unknown	laparotomy	LSO	EMA-CO	death
Yamamoto et al. [56], 2007	19	206,949	Non-Gestational	Unknown	Laparotomy	LSO	EMA-CO	1 year without disease evidence
Mishra and Cras-ta [57], 2008	25	>1000,000	Non-Gestational	Unknown	Laparotomy	TAH + BSO + omentum biopsy	unknown	lost follow-up
Heo et al. [4] 2004	12	20,257	Non-Gestational	IA	Laparoscopic	LSO + omentectomy + peritoneal biopsies	BEP	14 months with-out disease evidence
Santos et al. [8], 2009	10	206.949.70	Non-Gestational	IV	Laparotomy	TAH + BSO	none	death

TAH: Extended total abdominal hysterectomy; BSO: Bilateral salpingo-oophorectomy; LSO: Left salpingo-oophorectomy; RSO: Right salpingo-oophorectomy; EMA-CO: Etoposide, Methotrexate, Actinomycin-D, Cyclophosphamide, Vincristine; EMA-CE: Etoposide, Methotrexate, Actinomycin-D, Cisplatin, Etoposide; PVB: Cisplatin, Bleomycin, Vinblastine; Act-D: Actinomycin D; MAC: Methotrexate, Actinomycin-D, Cyclophosphamide; BEP: Bleomycin, Etoposide, Cisplatin; MTX: Methotrexate; VBP: Vinblastine, Bleomycin, cis-platinum; VAB-VI: Vinblastine, Bleomycin, Cisplatinum, Actinomycin D, Cyclophosphamide; AuBMT: Autologous bone marrow transplantation

Table 3: Gestational primary ovarian choriocarcinoma - literature review.

					Surgery			
Author / Reference / Year	**Age**	**Initial Beta HCG**	**Diagnotic**	**Stage**	**Via**	**Surgical Description**	**Chemotherapy regimen**	**Prognosis**
Lorigan et al. [13], 1996	41	151,500	Gestational		Laparotomy	TAH + BSO + omentectomy	BEP	3 months without disease evidence
Namba et al. [58], 2003	37	990,000	Gestational	unknown	Laparotomy	RSO	MAC	no evidence of disease -indeterminate time
Mood et al. [20], 2009	31	over 1000	Gestational	unknown	Laparotomy	RSO	EMA-CE	7 years without disease evidence
Aucouturier [11], 2003	43	37,260	unknown	unknown	Laparotomy	TAH + BSO + pelvic lymphadenectomy and para-aortic + omentectomy + peritoneal biopsies	BEP	1 year without disease evidence
Naniwadekar et al. [59], 2009	19	380,000	unknown	unknown	Laparotomy	TAH + BSO + omentectomy	EMA-CO	lost follow-up
Vautier et al. [60], 2004	32	535,000	unknown	IC	laparoscopy	LSO	BEP	1 year without disease evidence
Kar et al. [6], 2015	25	3,080	Gestational		Laparotomy	TAH + RSO	EMA-CO	Unknown

TAH: Extended total abdominal hysterectomy; BSO: Bilateral salpingo-oophorectomy; LSO: Left salpingo-oophorectomy; RSO: Right salpingo-oophorectomy; EMA-CO: Etoposide, Methotrexate, Actinomycin-D, Cyclophosphamide, Vincristine; EMA-CE: Etoposide, Methotrexate, Actinomycin-D, Cisplatin, Etoposide; PVB: Cisplatin, Bleomycin, Vinblastine; Act-D: Actinomycin D; MAC: Methotrexate, Actinomycin-D, Cyclophosphamide; BEP: Bleomycin, Etoposide, Cisplatin; MTX: Methotrexate; VBP: Vinblastine, Bleomycin, cis-platinum; VAB-VI: Vinblastine, Bleomycin,Cisplatinum, Actinomycin D, Cyclophosphamide; AuBMT: Autologous bone marrow transplantation

to the ovary, there is a scientific consensus to remove only the affected ovary, preserving the reproductive performance [4-9,12].

Follow-up and survillance

However, all medical-scientific literature advocates for combined treatment with surgery and adjuvant and/ or neoadjuvant chemotherapy and the response to the treatment is assessed by quarterly serial serum βhCG analysis for at least five years [1-5].

In a survey conducted at our institution (Table 4), the data show that from January 1991 to March 2016 the INCA recorded 33 cases of choriocarcinoma. Of these, only the object of this report is a primary ovarian choriocarcinoma.

The data from our institution reinforce the evidence of global medical literature about the rarity of Primary ovarian choriocarcinoma and also reinforce that early detection, assertive diagnosis and treatment as oncological urgency are key factors for the patient prognosis.

In our report, the initial approach to a young, tachypneic patient with high serum level of βHCG, abdominal scan showing ovarian tumor and thoracic scan revealing multiple pulmonary nodules with no association of pleural effusion, incited the assistant team to conduct the case as recommended by medical literature [1,8,12], with chemotherapy.

The rarity of Primary ovarian choriocarcinoma is

Table 4: Supplementary Material. Patients Admitted with Diagnosis of Choriocarcinoma at the Brazilian National do Câncer from Jan/1991 to Mar/2016

	N	%	Stage I (N)	Stage II (N)	Stage III (N)	Stage IV (N)
Ovary	1	3%	-	-	-	1
Uterus	32	97%	7	-	6	19
Age Group						
17–20	3	9%	-	-	2	1
21–40	26	79%	6	-	4	16
>40	4	12%	1	-	-	3
Diagnostic Year						
1991	1	3%	1	-	-	-
1993	1	3%	-	-	-	1
1996	1	3%	-	-	-	1
2001	2	6%	-	-	1	1
2002	1	3%	-	-	-	1
2004	1	3%	-	-	-	1
2005	4	12%	2	-	1	1
2006	3	9%	1	-	-	2
2007	4	12%	-	-	-	4
2008	2	6%	-	-	-	2
2009	2	6%	1	-	-	1
2010	1	3%	1	-	-	-
2011	2	6%	-	-	1	1
2012	1	3%	-	-	-	1
2013	3	9%	1	-	2	-
2014	3	9%	-	-	1	2
2015	1	3%	-	-	-	1

an incentive to research. Although there are studies, reports and case series, as well as bibliographic reviews, the medical literature still presents questions about the natural history of the disease and treatment regimens. Therefore, future research is fundamental for a more accurate understanding of the pathophysiology of this tumor type and for the emergence of new therapeutic perspectives with impact on survival.

Conflict of Interest

There is no one conflict of interest.

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